Offprint from
Advances and Technical Standards in Neurosurgery, Vol. 31
Edited by J.D. Pickard

Neurosurgical Concepts and Approaches for Orbital Tumours

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With 16 Figures

Contents

Abstract

Orbital tumours are lesions that appear within the orbital craniofacial borders. To this end, treatment of these tumours is assured by teams of different specialists. Furthermore, these pathologies are different in adults and in children. We have endeavoured, in this chapter, to highlight the specifically neurosurgical features of orbital tumours or, to be more precise, tumours affecting the posterior two thirds of the orbit and tumours originating in or intruding into the optic canal. The list of aetiologies is long. After recapitulating the main types of tumour (as well as those of most concern), we have also studied the different stages of surgery, namely approaches and reconstructions which we have illustrated at each stage by a tumour that, in our view, seemed emblematic of the problem in question: the lateral eyebrow approach for schwannoma and cavernous angioma, the transorbital subfrontal approach for optic nerve glioma, the pterional and orbital approaches for spheno-orbital meningioma, problems with reconstruction and with plexiform neurofibroma affecting the orbit and fibrous dysplasia of bone.

Keywords: Orbital tumours; optic nerve glioma; plexiform neurofibroma; lymphangioma; rhabdomyosarcoma; orbito sphenoidal meningioma; optic sheath meningioma; orbital vascular lesions; fibrous dysplasia; orbital reconstruction; neurofibromatosis.

Introduction, Definition of Subject and Limitations

The mass of literature relating to orbital tumours testifies to the significance and complexity of the subject. Diagnosis and treatment of these tumours is often a major challenge that is undertaken by a wide variety of different specialists: ophthalmologists, plastic surgeons, otorhinolaryngologists, neurosurgeons, paediatricians, oncologists etc. The neurosurgeon deals primarily with lesions to the posterior third of the orbit [4], or those that overlap onto the posterior third of the orbit and the contents of the skull. This anatomical description must be completed by the following, more restrictive information: Orbital Tumours (OT's) are neoplasms of the bony orbit and its content with the exception of the eyeball. Certain lesions that we have encountered include occupying processes that are not strictly tumours (for example, lymphangioma). We have therefore used the term 'neoplasm' intentionally in this context. To this end, we have excluded secondary tumours: those that stem from the face or from the eyeball and which affect the orbit at a later stage such as retinoblastoma in children.

Surgical anatomy is not the same as that of anatomy books. However, even if the latter are essential to understand and carry out this surgery, they are not particularly pertinent in the domain of neurosurgery since access to the orbital cone, the nerve and the optic canal is strictly reserved to the domain of neurosurgery. This study is neither a thesaurus nor an exhaustive description of orbital tumours that presents all the questions that all specialists might ask. Therefore, in order to avoid repeating fastidious lists of aetiologies our preferred approach was to proceed, step by step, from biopsy to surgical excision and examine OT's that correspond to the field of neurosurgery and which are representative of an indication, and of surgical techniques, that we will describe here.

Historical Perspective

The history of tumourous proptosis is often confused with the history of medicine and ophthalmology, but the first efforts at coherent classification are much more recent [8]. Antonio Scarpa in 1816, [67] offered the first clinical description of optic nerve (ON) tumours. Jean Cruveilhier (1835) [14] thought that meningioma should be considered as clinical entities that are entirely separate from tumours of the central nervous system. Von Graefe in 1854 [79] considered ON tumours to be entirely separate and attributed to them the first semiological criteria of diagnosis. It was not before Hudson discovered it in 1912 [32] that a clear distinction was made between optic nerve glioma and meningioma of the optic nerve sheath. Harvey Cushing [15] made the first distinction between primitive meningioma of the optic nerve sheath and intracranial meningioma that spreads to the orbit. The idea of approaching the orbit via the endocranial approach was successfully described and practised by Durante 1887 [22]. Surgery to the posterior part of the orbit was at its peak when neurosurgery first came into being less than a century ago. The problem of retracting the frontal lobe, which had always been considered as the main obstacle for this approach, was partially solved as early as 1913 when Frazier [26] suggesting the deposing of the orbital margin. The first neurosurgical principles for this were described by Naffziger [55]. According to this perspective, a pterional approach of the orbit was proposed by Hamby in 1964 [29]. Simultaneously, during the 1960's the first developments in orbito-cranial surgery of cranio-facial malformations came about with the findings of Converse, Tessier and Mustarde. On the basis of these technical principles, numerous works appeared that aimed to define the principles of reconstruction following excision [45, 2, 75]. It is still difficult, to this day, to dissociate the history of orbital surgery from the recent history of imaging: ultrasonography, CT scan using X-rays and MRI. Although it is not strictly the history of neurosurgery, we cannot leave aside IT imaging and its clinical and ther-

apeutic consequences. The first comparative description of orbital tumours examined by MRI and CT scan can be attributed to Li [42]. Today, imaging is not only a factor in diagnosis but also directly in the surgical procedures of intracranial neuronavigation as described by Shanno in 2001 [72] and possibly, in future, during surgery (intra operative MRI).

Aetiologies

Generalities

Aetiologies pertaining to orbital tumours are great in number, and often necessitate entirely different procedures. In order to avoid proposing surgery that may be, on occasions, useless (for example, rhabdomyosarcoma in children), or, inversely, to avoid deferring surgery that is imperative (in the event of orbital meningioma), it is necessary to detail the clinical and para-clinical characteristics of the principal aetiologies which relate to tumours. Adult and paediatric varieties of OTs constitute two clusters of distinctive histological entities (Table 1). Diagnosis of an orbital mass is based on clinical and evolutive elements as well as information obtained from modern imaging. Clinical examination has to be conducted with care: measurement, direction of proptosis, impairment of ocular motility, compressive optic neuropathy, age at the onset and unilateral or bilateral proptosis are important features. Benign tumours like dermoid cysts or haemangiomas grow slowly whereas rapid growth suggests a metastatic tumour or

Table 1. *Adult and Paediatric Varieties of OTs Constitute 2 Clusters of Distinctive Histological Entities*

OTs that occur primarily during childhood	OTs that occur primarily during adulthood
– Optic Nerve Glioma	– Orbital Meningioma
– Plexiform Neurofibroma	– Optic Sheath Meningioma
– Capillary Haemangioma	– Cavernous Haemangioma
– Lymphangioma	– Schwannoma (Neurilemoma)
– Rhabdomyosarcoma	– Fibrous Histiocytoma
– Dermoid Cyst	– Mucocele
– Teratoma	– Epithelial Tumours of the Lachrymal Gland
– Granulocytic Sarcoma (Chloroma)	
– Lymphoma	– Lymphoid Tumours
– Histiocytic Tumours	– Metastatic Tumours
– Metastatic Neuroblastoma	
– Fibrous dysplasia	

a rhabdomyosarcoma. Rapid bilateral proptosis associated with periorbital ecchymosis suggests a metastatic neuroblastoma. Pseudotumours of the orbit (idiopathic orbital inflammation) can go through growth spurts; however they manifest themselves with pain, impairment of ocular motility and hyperthermia. Fundi and visual acuity should be checked. General examination focuses especially on cutaneous abnormalities: café au lait spots in NF1, haemangioma that often involves the skin of the eyelid. Meningioma of the orbit is frequently associated with a palpable mass in the external temporal fossa. Ultrasonography is available in an office setting but is limited in its ability to accurately measure the size and infiltrating properties of the lesion. The depth to which it can penetrate in the orbit is limited (20 mm). This leaves the posterior third of the orbit inaccessible to this kind of imaging. Most of the time standard X-rays, CT and MRI permit diagnosis. In children, CT scans and MRI sometimes require the use of general anaesthesia in order to be properly performed.

Main Orbital Tumours Occurring in Adulthood

Orbitosphenoidal Meningioma (Fig. 1)

Orbito sphenoidal meningiomas represent 18% of intracranial meningiomas. They combine, with varying degrees, the following: invasion of the great wing of the sphenoid, invasion of the lesser wing of the sphenoid that may surround the optic canal and the anterior clinoid process, the adjacent frontotemporal dura, the dura of the superior orbital fissure with, on occasions a large bud in the orbit that is often in an extraconical position, and, sometimes, the cavernous sinus (Fig. 1 a, b). Proptosis (90% of cases) is clinically dominant. Other signs are: reduction of visual acuity (50% of cases), reduced field of vision, the oculomotor nerves may be affected but this is more rare, and the trigeminal nerve may be affected on very rare occasions. Analysis of the tumourous extension is conducted by means of a CT scan using the bone window level that is effective for visualising intraosseous extensions, and imaging by MRI to show intraorbital extensions, periorbital extensions and those affecting the area of the cavernous sinus. The inevitable progression towards blindness of the affected eye justifies a surgical indication [18]. The frontotemporal pterional approach is sometimes completed by deposing the orbito zygomatic arch. We never practise this according to this indication, but we do think it is necessary to systematically open the optic canal after having drilled the anterior clinoid process. The lateral wall of the orbit, opened wide, is not reconstructed in order to relieve and treat the proptosis. Attempts at excision within the cavernous sinus are deleterious for the oculomotor nerves and it seems

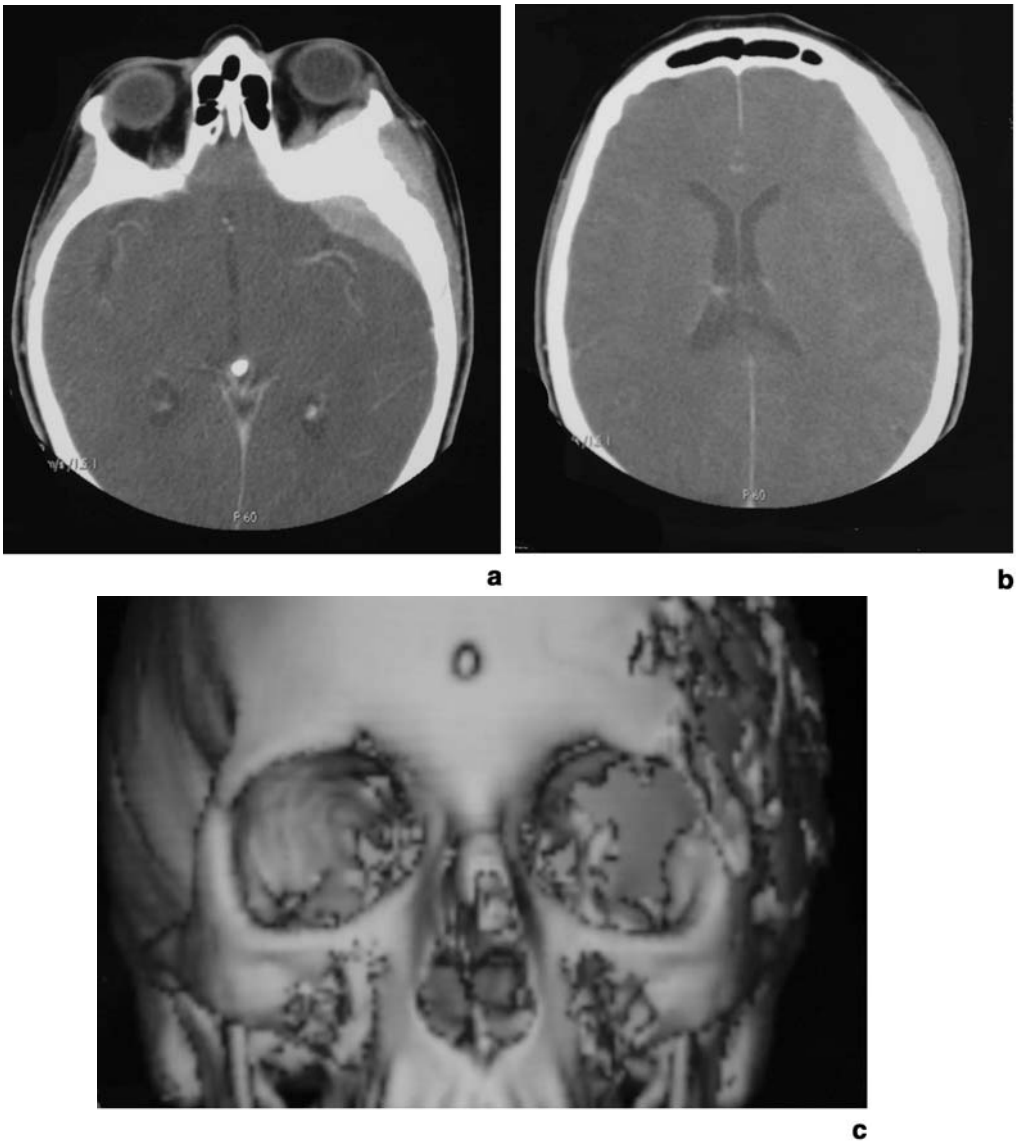


Fig. 1. Orbito Sphenoidal Meningioma: (a, b) Computed Tomography (*CT*) with iodine contrast: left sphéno-orbital meningioma involving the orbit, the vault and the soft tissues. (c–d) Frontal (c) and sagittal (d) views of 3D post-operative CT. Reconstruction with split ribs of the lateral orbital margin (d). Note the wide surgical opening of the superior orbital fissure (c)

Fig. 1 (*continued*)

preferable to complete a partial excision at this level by radiosurgery or conformational radiotherapy. The results are often satisfying: reduction of proptosis and improvement in visual acuity [43]. Surgery can be complicated by the deterioration of visual acuity following surgery, especially if the optic nerve was previously affected [27]; post-surgical ptosis that is, in most cases, transient, is generally the result of manipulation of the levator palpebrae muscle/superior rectus muscle complex. Orbital-sphenoidal meningiomas tend to spread across the foramen, canals, fissures, periorbit, dura mater and the bone, thus giving these tumours a microscopic dimension that cannot be taken into account easily during the operation, and as a result the term 'complete excision' should be used with extreme care [49]. Partial excision alone can explain recurrences: from 34 to 54% of cases according to [51, 1], based on a follow-up study of 5 or 10 years. In the event of excision that is considered incomplete due to extension of the cavernous sinus, conformational radiotherapy or post-surgical radiosurgery is preferred instead of approaching the cavernous sinus directly: with a follow up of 8 years, 88% of patients undergoing surgery plus radiotherapy do not experience recurrence compared with 48% in the case of isolated surgery [49].

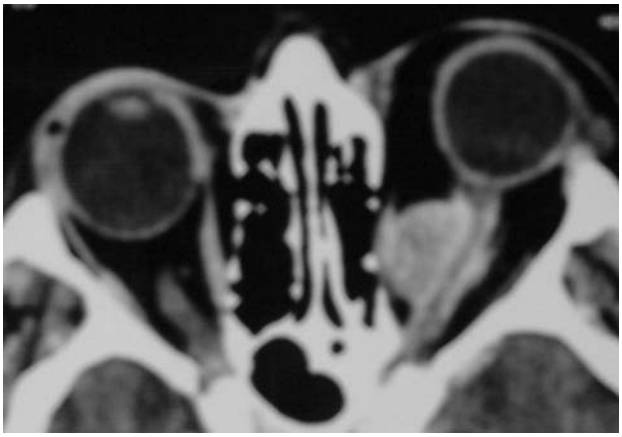


Fig. 2. Optic Nerve Sheath Meningioma: CT with iodine contrast: left optic sheath meningioma. This axial view shows the meningioma stems from the nerve sheath itself

Optic Sheath Meningioma (Fig. 2)

Second only to gliomas, this type of optic nerve tumour is the most frequent. They represent between 1 and 2% of intracranial meningiomas [23]. Efforts at excision of optic sheath meningioma are generally complicated by post-surgical amaurosis due to the problem of finding a suitable cleavage between the optic nerve and its tumour on the one hand, and vascular traumatism provoked by dissection around the nerve on the other [16, 71]. If visual acuity is preserved, radiotherapy that is fractionated under stereotactic conditions could, today constitute a good option for therapy: with an average period of 89 weeks of monitoring, Andrews *et al.* remarked that, for 92% of cases, visual acuity had been preserved compared with only 16% for the group with a single clinical follow-up [3]. When visual acuity is no longer functionally useful, a neurosurgical operation using the intraconical approach avoids the intracranial tumourous extension via the optic chiasm [56].

Cavernous Haemangioma

Cavernous haemangioma is mostly found in women aged between 40 and 50 [57]. It usually manifests itself as a proptosis and is distinct in that it appears progressively. It is indolent and reducible, thus revealing its vascular nature. When the cavernous haemangioma is in contact with the optic nerve, it can cause a reduction in visual acuity or diminish the field of vision. Echography is particularly useful here as it highlights the cystic nature of the lesion. Excision of cavernous haemangiomas is simple due

to their non-infiltrating and encapsulating nature. The only surgical complication here would be adhesion to the optic nerve. The lateral orbital approach is the most suitable indication when the orbital cavernomas is located in the lateral part of the intraconal space. When the optic canal or the superior orbital fissure are concerned, an pterional extra/intradural approach should be preferred [69].

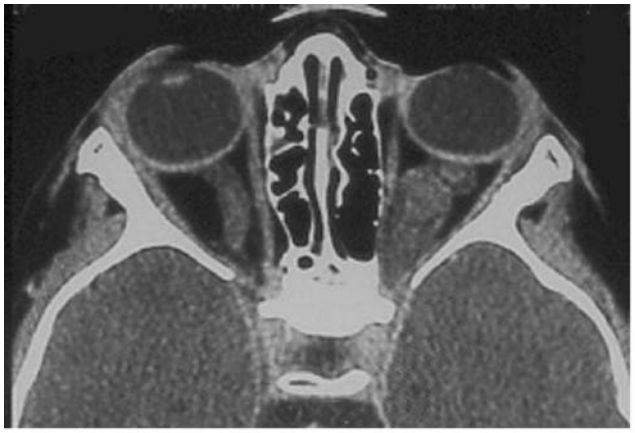
Schwannoma (Neurilemoma)

Schwannoma are nervous benign tumours that develop at the expense of the cells of the Schwann sheath. The name 'schwannoma' is attributed to Masson (1932) and the term neurilemoma to Stout (1935). It was not until 30 years later, when electronic microscopes were invented, that the original cell of this tumour was associated with Schwann cells. These tumours occur in young adults and are rarely associated with NF1. They represent 1% of orbital tumours [39, 65]. Orbital dystopia with proptosis is the revealing factor. They seem to develop more readily at the expense of the nervous fibres of the frontal nerve [25]. They develop mostly in the superior part of the orbit, touching the roof in an extra-conical or intra-conical position [60]. The tumour may be extirpated using a lateral intraorbital approach by debulking when it is large [70].

Main Orbital Tumours Occurring in Children [61, 68]

Optic Nerve Glioma (ONG) (Fig. 3)

Tumours of the optic nerve are rare, representing only 4% of all gliomas and 2% of all intracranial tumours. The peak frequency is between 2 and 8 years at a ratio of 3 girls to 2 boys. 75% of ONG's are diagnosed before the age of 10 years and 90% before the age of 20. They can be isolated or associated with NF1. They are pathognomonic of this disease when they are bilateral. 10% to 38% of patients suffering from an ONG have NF1, whereas 15% to 40% of those suffering from NF1's have optic pathway glioma. 50% to 85% of all optic pathway gliomas involve the optic chiasm or the hypothalamus. 80% of patients with an NF1 have an ONG that is limited to the optic nerve. ONG is the second most common imaging abnormality in patients with NF1 following the T2 hyperintensity seen in the basal ganglia, internal capsule, brain stem and cerebellum [31]. Tumours of the optic nerve can be classified according to their supposed point of origin: those situated on the optic disk which are intraocular, those situated in the orbit which are intraocular and constitute the subject of this study and those which invade the intracranial part of the optic pathways. In children, these tumours are usually quiescent and thus pose the problem of their



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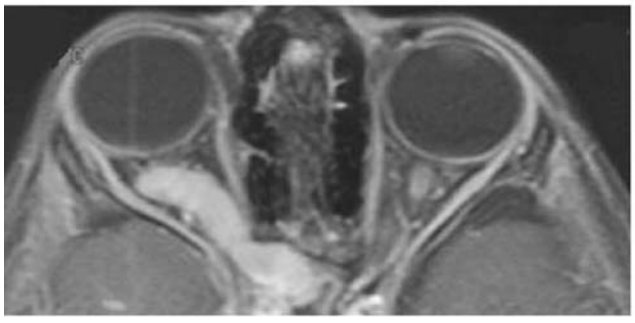

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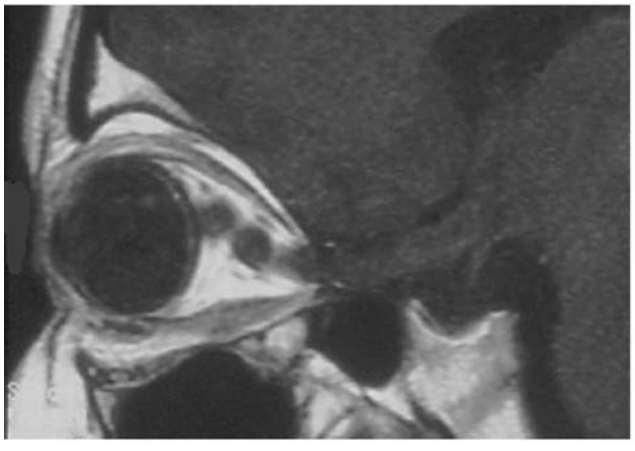

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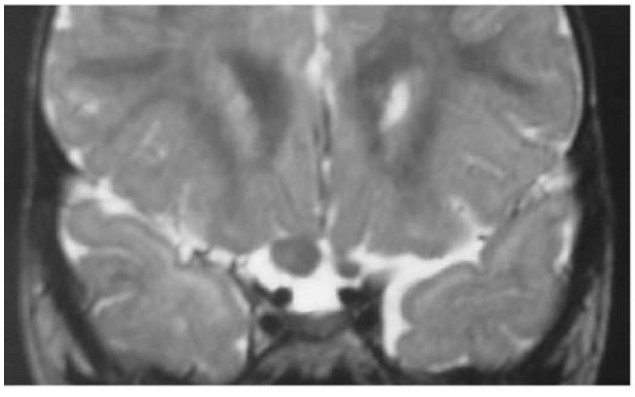

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hypothetical tumourous nature. In rare cases these tumours may present evolutive growth spurts before the age of 2 or during adolescence. Spontaneous regressions are recognised. When they evolve in an aggressive way they can spread forwards and aggravate the proptosis, or backwards thus invading the optic chiasm via the optic canal (Fig. 3 b). Their histological nature likens them to pilocytic juvenile astrocytoma. Intraorbital gliomas are revealed by a reduction in visual acuity. There are no oculomotor palsies. Proptosis occurs later along with optic atrophy. Papille oedema is rare. A reduction in visual acuity in a patient suffering from NF1 is pathognomonic of an ONG. The clinical sign of an ONG in children is a reduction in visual acuity that cannot be corrected. When the visual field is affected, the ONG manifests itself as a central or paracentral field defect [31, 19, 21]. Before the invention of modern imaging techniques diagnosis was based on radiography of the orbit and optic canal that looked for a widening of the optic canal of more than 5 mm, and arteriography showing the widening of the loop of the ophthalmic artery as it passed around the optic nerve. Today CT scans permit effective analysis of osseous modifications but both with and without a contrasting agent, its use is limited in order to study the optic nerve in detail (Fig. 3). MRI is the most effective technique for studying modifications to the optic nerve (Fig. 3 d). The examination uses fat suppression techniques and a set of oblique sagittal images which are aligned parallel to the orbital course of the optic nerve (Fig. 3 c), from the globe to the chiasm [31]. These tumours have a low growth potential, therefore vision is affected at a later stage. Furthermore, the treatment is conservative and only requires annual evaluation by MRI. Tumours that grow rapidly are problematic for surgical indication. If visual acuity is preserved, surgical resection of the tumour would constitute mutilation. If the surgeon is confronted with a tumour that is clearly surgically, or radiologically evolutive, it is better to propose either chemotherapy or radiotherapy in the first instance rather than surgical excision. In this case, it is clear that repeated MR must be performed. In effect, surgical excision should take place before the tumourous extension spreads backwards in the direction of the optic chiasm. In the event of straightforward clinical and radiological surveillance, there is no question of a biopsy. The decision to carry out either radiotherapy or chemotherapy in the case of rapid growth may raise the question of surgical biopsy: on the one hand,

Fig. 3. Optic Nerve Glioma: (a) CT: left optic nerve glioma circumscribed into the orbit. (b) T1 weighted MRI: right optic nerve glioma spreading backward to the chiasm. (c) T1 weighted MRI: optic nerve glioma on an oblique sagittal image which is aligned parallel to the orbital course of the optic nerve from the globe to the chiasm. (d) T2 weighted MRI: frontal view of a right optic nerve glioma

the appearance of the lesion when examined by MRI is characteristic, but on the other hand biopsy on the optic nerve constitutes a functional risk that is sufficiently great to dismiss this option. Indication of a surgical resection of an ONG is justified if the tumour remains anterior to the optic chiasm, and may be completely resected if it presents clinical or radiological signs of evolution that suggest possible extensions to the optic chiasm, and if visual acuity is severely impaired or not functional (amblyopia). The techniques for such a resection include an intraorbital and intracranial approach of the optic nerve.

Plexiform Neurofibromas (Fig. 4)

It is common practice to separate neurofibromas into 4 groups [11]: plexiform, diffuse, circumscribed and postamputation. Plexiform neurofibromas are tumours which occur in children, and the circumscribed types are schwannoma. This type of tumour, the description of which is attributed to Von Recklinghausen in his 1882 study, seems to have existed for much longer as Huson identified in 1994 [33] when he related an illustration by Aldrovandi that dated from 1642. Ophthalmic-orbital deformity, generally unilateral, can be associated with what is obviously dystopia. Such a location is quite rare, estimated at 1% to 7% of patients with an NF1 [34, 41, 52]. Plexiform neurofibromas affect the eyelids, and the initial clinical manifestation is usually a visible swelling of the superior eyelid, thus giving it a hypertrophic appearance (Fig. 4 a). On palpation these nodules give the impression of "a bag of worms". They provoke intraocular hypertension that is responsible for proptosis in children, or glaucoma. Nonetheless, the most evocative sign is pulsatile proptosis [52, 66]. This is secondary to an intraorbital hernia of the temporal or frontal lobe, made possible by agenesis of the great wing of the sphenoid, enlargement of the superior orbital fissure and defects of the orbital roof. The aspect of facial asymmetry and proptosis stems from the imbalance between the contents of the orbit and the orbital container [10]. In effect, if the tumourous growth affects either one or the other of the parts, it is the ocular globe that suffers the constraint. During adolescence the tumour extends to the forehead, temple and superior orbit leading to downward displacement of the globe. Osseous defects of the affected part of the orbit are the result of lysis of the posterior orbit, but also of the constrained growth of the anterior orbit by hyper pressure. They associate partial or total absence of the great wing of the sphenoid and malar hypoplasia (Fig. 4 b) [6, 41, 37, 38]. CT and MR imaging show nodular lesions with significant contrast enhancement, with irregular limits, thus giving it an infiltrating appearance. Treatment of plexiform neurofibromas is not satisfactory, as they infiltrate the normal anatomical structures such as the oculomotor muscles and the extraocular

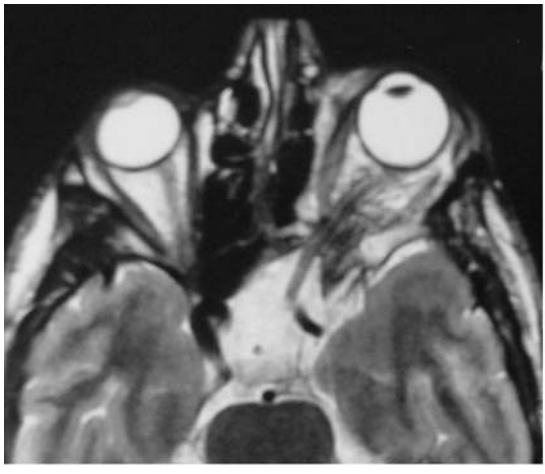

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Fig. 4. Plexiform Neurofibroma: (a) plexiform neurofibroma in an adolescent: note the "S" shaped eyelid and the extension to the face. (b) CT (axial view): the dysplasia of the sphenoid wings is associated with a plexiform neurofibroma

muscles. They have a high rate of recurrence and may require numerous operations in order to obtain a satisfactory aesthetic result. In addition to difficult and often incomplete resections, due to their infiltrating nature, they pose the problem of osseous reconstruction of the orbital roof defect that has deteriorated due to orbital-sphenoid dysplasia. Bone grafts are often difficult in terms of finding a suitable resting point on an affected bone that has been destroyed or eroded by the plexiform neurofibroma. Surgery is therefore complex, multidisciplinary, staggered over time and is rarely neither aesthetically nor functionally satisfactory.

Orbital Vascular Lesions [5]

Capillary Haemangioma. Capillary haemangioma is the vascular lesion that occurs most frequently in infants. It may be present at birth or appear a short while after. It is situated in the deep or superficial areas of the orbit. There are, on occasions no cutaneous manifestations. The tumour tends to affect the superomedial part of the orbit. It can invade the entire orbit. They occur most often in females. They generally grow rapidly during the first 6 months of the infant's life and then begin to decrease in volume. Cutaneous localisations elsewhere on the body can be found in 30% of cases [21]. 60% disappear completely by the age of 4 years and 76% by the age of 7. The size of the tumourous mass can sometimes displace the ocular globe and hinder vision. Imaging is particularly important if the lesion is deep, and has no clinical cutaneous manifestation. Its internal architecture is characteristic: irregular mass, well-defined and enhancing with contrast. A differential diagnosis with a rhabdomyosarcoma, a metastatic neuroblastoma or an orbital lymphangioma is, in theory, simple. Their treatment depends essentially on their size and the way in which they hinder vision. The risk of amblyopia should be treated with corrective glasses, taking care to hide the unaffected eye. The treatments proposed are: corticosteroids by general administration or local injection or interferon (inhibitors of angiogenesis). Surgery must always take into account the natural evolution which is often spontaneously satisfying.

Orbital Lymphangiomas (OLs) or Venous Lymphatic Malformation (Fig. 5). It is likely that orbital lymphangiomas are of congenital origin [40]. These malformations have a malformed component that is a mix of venous and lymphatic. They appear at the level of the head and neck, spreading to the orbit. They are diagnosed at birth or in young infants (diagnosis is generally performed at the age of 6 years). They are slow to evolve but very invasive. Rapid growth spurts relate to hypertrophy of the lymphatic tissue that is provoked by infections of the ENT sphere, the upper respiratory

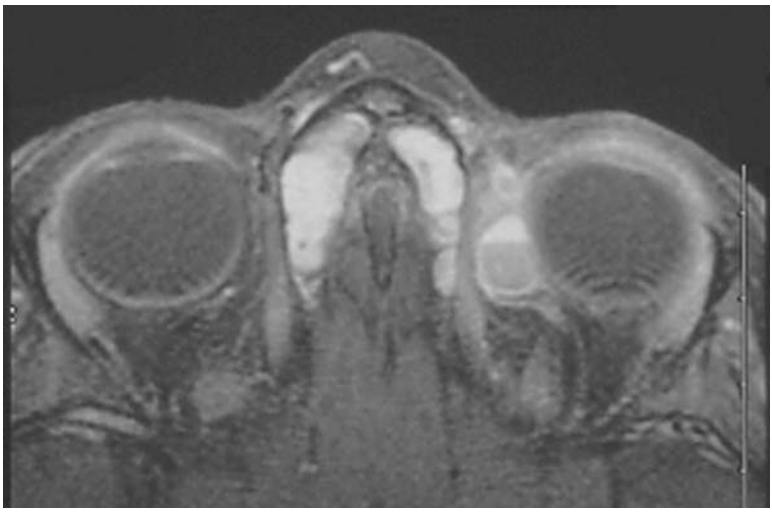
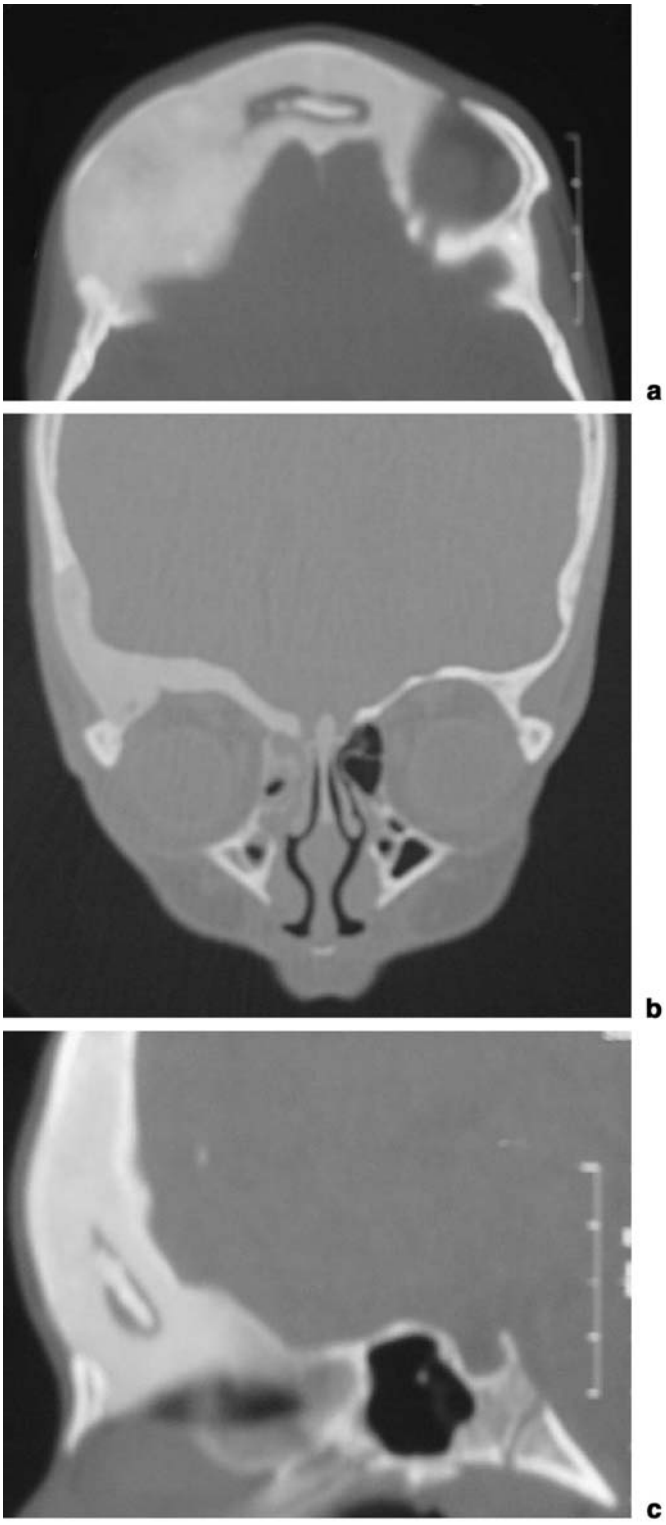


Fig. 5. Lymphangioma: Typical "chocolate cyst" in a left orbital lymphangioma

tracts or the face. In other locations, there are haemorrhagic phenomena that produce cysts, typically known as "chocolate cysts". Imaging allows for a differentiation between fleshy parts and cystic areas. OLs are not encapsulated and, when positioned at the level of the orbit, they infiltrate the fat, orbital septa, nerves and muscles. These characteristics make complete excision impossible. Surgery should therefore be limited to decompression in order to relieve troubles related to the optic nerve or oculomotor nerves, or reduce a proptosis that is of worrying proportions. Such decompression implies either aspiration of a cyst or debulking of the tumour (which explains why effective pre-surgical imaging is essential). It is on the basis of these principles that a limited approach can be proposed.

Primitive Bone Tumours

Fibrous Dysplasia (Fig. 6.a–c). Fibrous dysplasia is a rare, benign condition that causes the replacement of normal bone with fibro-cellular tissue. Islands of normal bone are dispersed within this fibro-cellular tissue. In children, however, there may be growth spurts that can provoke proptosis. Initially described by Von Recklinghausen in 1891 [80], fibrous dysplasia represents 2.5% of all bone tumours [24]. This pathology tends to affect the ribs, the tibia and the craniofacial bones. It may appear as one of two forms: a focal presentation which is more frequent but for which a craniofacial localisation is rare; 10% to 27% of cases according to Munro, [53, 54], and a multifocal form that is characterised by the impairment of at least two non-contiguous osseous structures. Albright's syndrome is a par-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ticular type that combines fibrous dysplasia, sexual precocity, cutaneous café au lait spots, premature bone maturation and hyper parathyroidism. Orbital location is, in the majority of cases, isolated. Consequently the orbit is directly implicated at different levels, more readily in its superior hemi-circumference. In most cases, its initial manifestation is of aesthetic importance due to bone deformation. It appears as facial asymmetry. It may also lead to functional repercussions: reduction of visual acuity, epiphora caused by obstruction of the lachrymal canal in the event of ethmoidal involvement. From a vertical view, the ocular globe appears to be lowered and, from a horizontal view, the displacement can be lateral or medial. Imaging can be used for visualising a homogenous lysis or an osseous sclerosis, the two aspects mentioned can closely intermingle in various ways. Fibrous dysplasia of the orbit usually manifests itself during the first ten years of a person's life. Its growth during puberty is slow. Once this period has passed, evolution of dysplasia is extremely variable, ranging from a steady growth to stabilisation or, in exceptional cases, regression [76]. Its evolution is dominated by risk of damage to vision by compression of the optic nerve during its intra-osseous course. This justifies regular monitoring of acuity, both of the visual field and also the perception of colour. Risk of malignant transformation appears to be quite low and is estimated at between 0.5% and 4%. This potential evolution represents a greater risk for sufferers of Albright's syndrome. This evolution usually provokes pain and a rapid increase in the volume of the tumour. This has been observed after radiation of the dysplastic process [12, 13]. The surgical indication is entirely subject to the reduction in visual acuity, or to a severe narrowing of the optic canal that can be clearly radiologically demonstrated [12]. Aesthetic damage should justify the surgical indication on the condition that it does not infer a significant functional risk.

Osteosarcoma [19, 21]. Orbit osteosarcoma following radiation therapy for bilateral retinoblastoma is not rare. The occurrence of these tumours is due partly to radiation therapy in childhood, and partly to genetic abnormalities within chromosome 13. 30% of children who underwent radiation therapy for bilateral retinoblastoma harbouring chromosome 13 abnormalities will develop radio-induced sarcoma. Prognosis is poor and the five year survival rate does not exceed 30 to 40%.

Fibrosarcoma. Congenital, infantile, juvenile and adult forms of fibrosarcoma are recognized. Congenital and infantile fibrosarcomas are rare,

Fig. 6. Fibrous Dysplasia (*FD*) of the roof of the right orbit: (a–b) CT axial and frontal views of FD of the right orbit; (c) sagittal reconstruction

grow rapidly and have a low incidence of metastasis (less than 8%). Juvenile fibrosarcoma of the orbit has effective prognosis in children under ten years old when treated by radical local excision. Fibrosarcoma of the orbit is second only to osteosarcoma, following radiation therapy for retinoblastoma.

Histiocytic Tumours. Histiocytosis includes three groups of diseases of increasing gravity and juvenile xantho-granuloma.

Eosinophilic Granuloma: Eosinophilic granuloma has a rapid onset. It looks like an inflammatory pseudo-tumour of the orbit with pain and supero-temporal swelling. Proptosis is unilateral. Blood cell count reveals a hypereosinophily and biopsy a reticulo-histiocytic hyperplasia.

Hand-Shuller-Christian Disease: Is characterized by a polyuro, polydipsic syndrome (insipid diabetes), bilateral proptosis, and defects on plain films of the skull.

Letterer-Siwe Disease: Represents the acute presence of histiocytosis in infants. Patients suffer from generalized malaise, hyperthermia and axial purpura, which may affect the eyelids.

The first two diseases have a variable response to corticoids and chemotherapy whereas Letterer-Siwe disease evolves dramatically and prognosis is poor.

Juvenile Xantho-Granuloma: Is often classified with this group of histiocytic diseases. Only children under 1 year old are concerned. Clinical presentation combines cutaneous papules with ophthalmologic manifestations: anterior uveitis and secondary glaucoma. Biopsy shows an abnormal proliferation of histiocytes, lymphocytes, eosinophilic and Touton's giant cells, which are the hallmark of the disease.

Sarcomas [19, 21]

Rhabdomyosarcoma (Fig. 7). Rhabdomyosarcoma are tumours frequently found in children, an orbital position being the most common. They are the most frequent of all malignant tumours of the orbit in children. They stem from rectus muscles, although it is extremely difficult to ascertain their true origin. The average age for diagnosis is 7 years and upwards, and the tumours predominate in males (3 boys to 1 girl). Natural evolution is serious: local extension towards the nasal fossa, para-nasal and parameningeal areas and general extension with pulmonary and hepatic metastasis. Tumourous growth and proptosis are rapid, thus necessitating emergency treatment. The proptosis often appears with inflammations. CT and

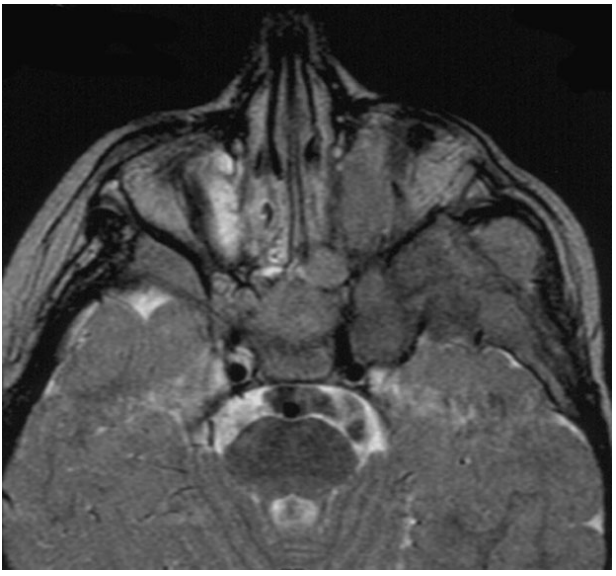


Fig. 7. Rhabdomyosacoma: Rhabdomyosarcoma of the left orbit. Dura and bone are concerned

MRI reveal the involvement of the intra-conical and extra-conical sections of the orbit. The most common starting point is at the supero-internal quadrant. The mass is ill-defined and has irregular limits. Contrast enhancement is low. Extension and invasion of the neighbouring osseous structures is very common, particularly the para-sinusal and para-meningeal areas. It is firstly necessary to confirm diagnosis by carrying out either surgical biopsy or biopsy using a needle. There are two kinds of histological rhabdomyosarcoma entities found in children: the embryonal variety and the alveolar variety. Prognosis is more positive for the alveolar variety than the embryonal variety. Currently, the preferred treatment is chemotherapy followed by radiotherapy. Surgery may be considered after the chemotherapy and radiotherapy, in order to confirm or invalidate the presence of tumourous residue on an image showing suspect residue after treatment. The global survival rate at the age of 5 years is 90%.

Fibromatosis. Is a benign but locally aggressive fibroblastic lesion that can be clinically and pathologically classified between fibrosis and fibrosarcoma. It affects mainly boys at the rate of 2 to 1 girl. It may be solitary (73% of cases), multicentric or generalized.

Nodular Fascitis. Is a common fibrous tumour found elsewhere in the limb and trunk in adults, but head and neck localization is more common

among infants and children. The tumour is well circumscribed. It is best treated by complete excision.

Leukaemia and Orbital Lymphoproliferative Disorders

Leukaemia. Occurs at any age and represents one of the most common forms of malignancy in childhood. Acute Lymphoblastic Leukaemia (ALL) accounts for 80% of all cases in children. About 80% arise from B-cell lineage and 20% from T-cell. Acute Myeloid Leukaemia (AML) is more common in adults, although it represents 20% of cases in children. There are many manifestations of orbital and ocular leukaemia. Orbital leukaemia is rare in ALL, but more common in AML. Such involvement in AML occurs in the form of *granulocytic sarcoma*, characterized by an infiltration of immature cells of myeloid (granulocytic series). This tumour is also known under the term of *chloroma* because of the myeloperoxydase within the mass, producing the green hue observed during microscopic examination. This orbital mass may precede the blood and bone marrow findings of AML or develop after the diagnosis. The most common ocular manifestation is retinal haemorrhage, which occurs when the patient is anemic or thrombocytopenic. Leukaemic infiltration of the optic nerve is an emergency because vision may deteriorate rapidly. Radiation therapy posterior to the globe and orbit on an emergent basis may be necessary to preserve vision. Leukaemic cells may also infiltrate the iris and the anterior chamber as present as iritis. These infiltrations may cause a secondary glaucoma. *Granulocytic Sarcoma* is a chloroma with bone erosion in the absence of blood involvement. This may be the first manifestation of AML. This lesion affects the subperiosteal space, usually the lateral wall of the orbit with extension in the temporal fossa or medial wall of the orbit with extension to the ethmoidal air cells and cribriform plate.

Lymphomas [78]. Extranodal manifestations of Non Hodgkin Lymphoma are common, but the incidence of orbital localization is low. There is usually an insidious presentation. This characteristic helps to differentiate lymphoma from orbital pseudo-tumours that progress rapidly over days. Burkitt's lymphoma (BL) is a disease due to the Epstein-Barr virus. BL is much more frequent in Tropical Africa. In 60% of cases, BL appears as a tumour of the maxilla which then affects the orbit, leading to a huge proptosis.

Metastatic Tumours of the Orbit

Metastatic Neuroblastoma (NB). Is a malignant tumour of the sympathetic nervous system and the most frequent orbital metastasis to occur during

childhood. Most of the time, primitive tumours are diagnosed prior to the metastasis. However, this diagnosis has to be kept in mind because orbital metastasis reveals the NB in 10% of cases and necessitates systematic abdominal palpation and echography. The primitive tumour usually appears in the medullo-suprarenal glands or other retroperitoneal structures. The sites of origin may be cervical, mediastinal and pelvic sympathetic system. These orbital metastases lead to unilateral or bilateral proptosis which is quite characteristic when they manifest ecchymosis of the eyelids (Hutchinson's syndrome). 90% of cases have an increased level of Vanilyl-Mandelic Acid in the urine because of cathecholamine secretion in the tumour.

Ewing's Sarcoma. Is a bone tumour, which may give rise to orbital metastasis.

Epidermoid Cysts (ECs) (Fig. 8)

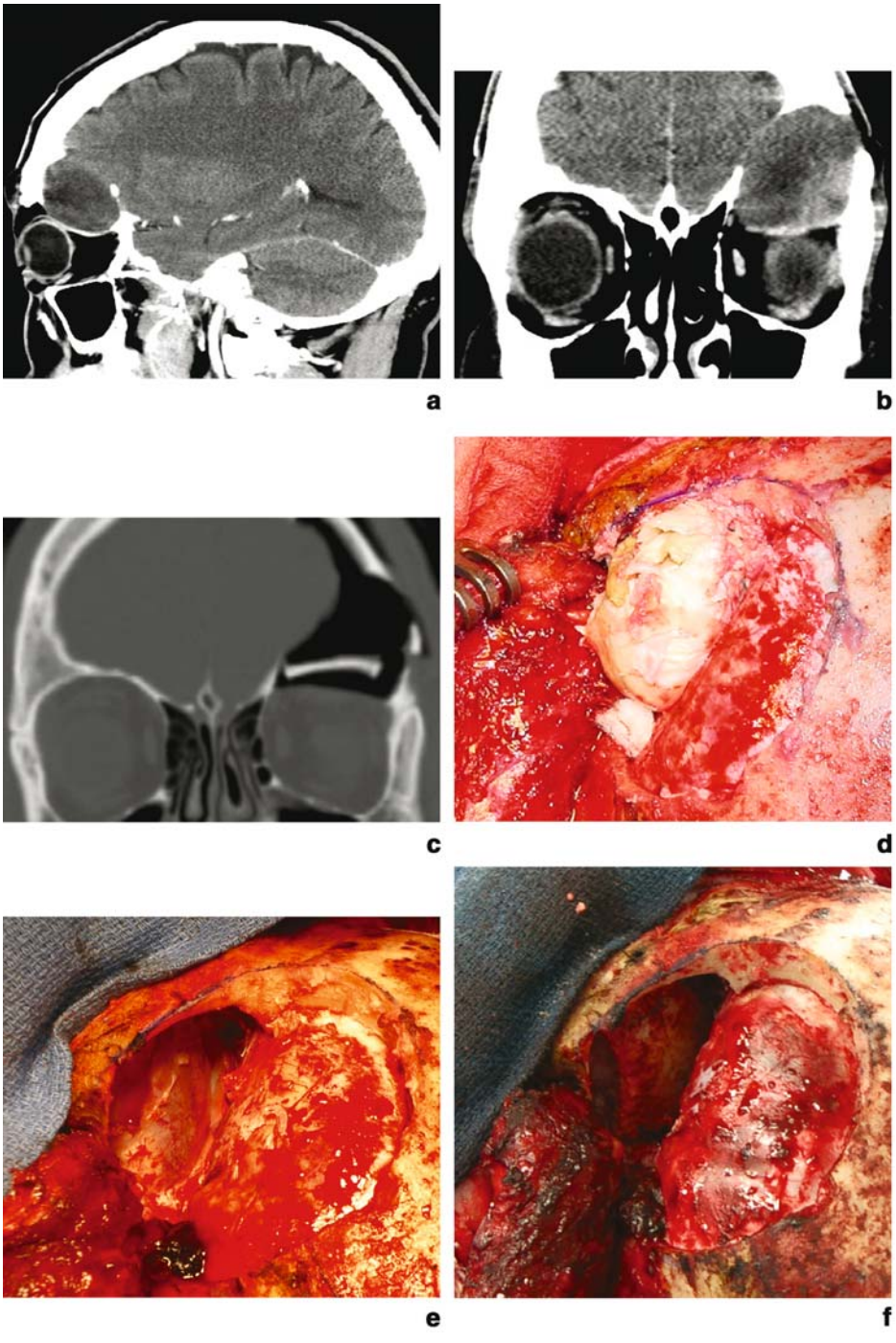
ECs tend to appear either as asymptomatic superficial lesions in children or as complicated deep lesions in adolescents and adults. Superficial ECs are more frequently lateral than medial. An eyebrow location is common. Superficial lesions can be dealt with by a direct, uncomplicated surgical approach. Surgical procedure must include an *en bloc* excision to avoid rupturing the cyst and to avoid inflammatory reaction in the post-operative courses. The deep lesions, in contrast, are frequently extensive and difficult to remove (Fig. 8 d–f), requiring careful pre-operative planning (Fig. 8 a–c) [73].

Teratoma (Fig. 9)

Teratoma is a cystic, benign, and congenital tumour. It is visible at birth and appears as a huge, impressive proptosis. It is often difficult to know where the eyeball is. CT scans show a multilobulated mass involving the neighboring anatomical structures (brain, bony orbit, etc.). Histological findings show a cyst surrounding the epidermis, gastro-intestinal mucosa or respiratory epithelium. On rare occasions, certain cases are more distinct, making up parts of a foetal body. The growth of these tumours is very rapid, extensive, and life threatening. They must therefore be removed as completely as possible even if surgery in neonates may be hazardous.

Surgical Approaches

As a general rule, it is necessary to ask the question why, for orbital tumours an orbital approach is more readily practised in the domain of neurosurgery than in the domain of ophthalmology or plastic surgery.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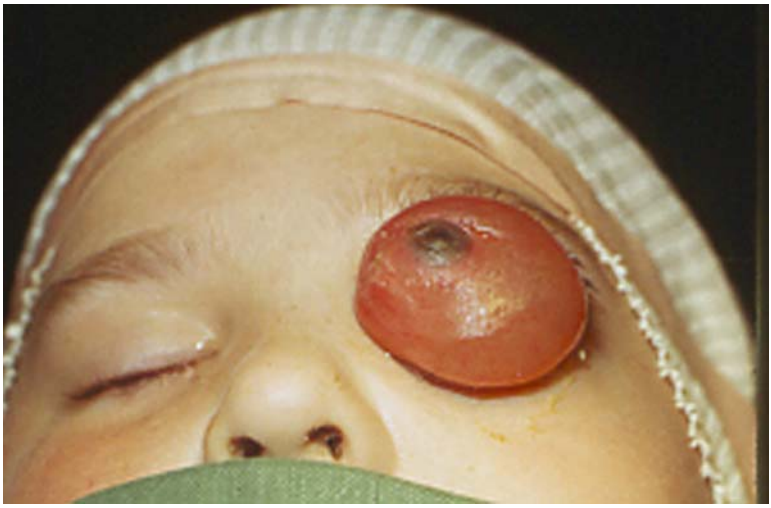
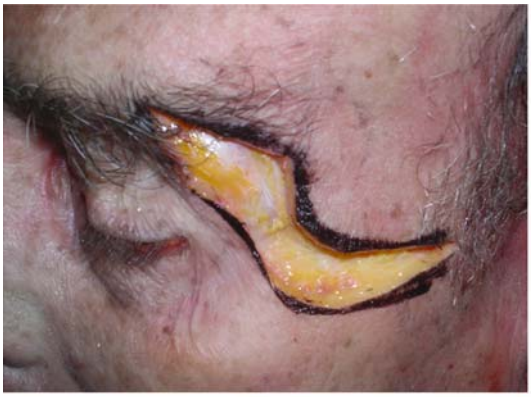


Fig. 9. Teratoma

There appear to be two circumstances that justify this decision: it is preferable to choose a neurosurgical approach in the case of lesions to the two posterior thirds of the orbit and/or in the event that it is necessary to check the excision of the tumour as well as that of reconstruction via an epidural or intra-dural approach. There are 3 surgical approaches that correspond to surgery of increasing difficulty which also suffers post-operative complications that become increasingly serious: the strictly intraorbital approach, the epidural approach and the intra-dural approach. In order to guarantee maximum safety during surgery, a difficult stage of the surgical approach should give the opportunity to check the approach to which it is contiguous: the intra-orbital approach should allow for epidural monitoring, and the epidural approach should allow for intra-dural monitoring. The intradural approach allows for monitoring all three but presents more deleterious side effects, and should therefore be reserved for accessing the anatomical structures such as the optic nerve, or for intra-dural tumourous extensions. The different approaches described here take place in microsurgical conditions whenever the orbit or the interior of the dura are operated upon.

Fig. 8. Epidermoid cyst of the left orbit (*EC*): (a–b) CT (a: sagittal reconstruction, b: frontal view) of EC of the left orbit. (c) CT (frontal view) reconstruction of the roof of the orbit with split bone flap. (d–f) Intra operative views (left sub frontal approach). (d) EC becomes visible beneath the dura, through the orbital roof. (e) EC is excised, note the large defect in the orbital roof. (f) the split bone flap is inlaid to replace the orbital roof



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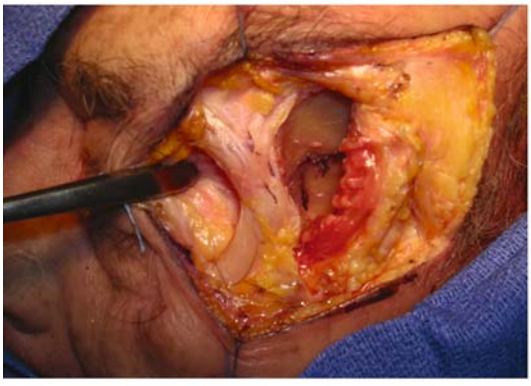

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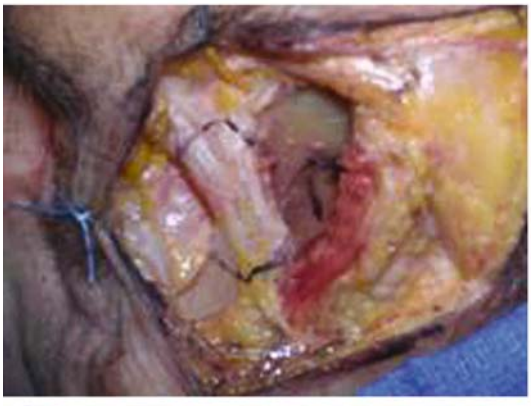

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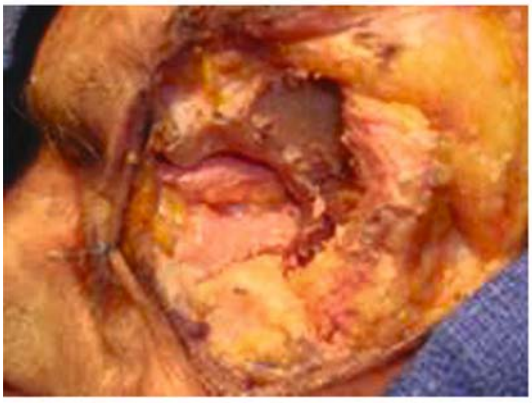

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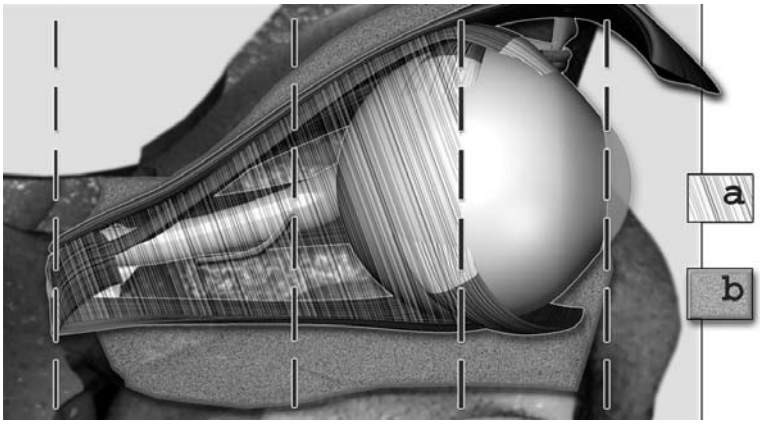


Fig. 11. The two posterior third of the orbit

The Lateral Intra-Orbital Approach (Fig. 10)

Surgical approach of the orbit and, in particular when concerning the two anterior thirds [4] can be adequately addressed by the classic ophthalmologic approaches, whatever variety or variant they may be. On the contrary, neurosurgical approaches that allow access to the posterior third of the orbit are more aggressive and can, on occasions represent a risk for the frontal lobe during retraction operations. Furthermore, the lateral approach as described by Krönlein is of particular interest since it associates the benefits of a trans browsal and those of an orbitotomy of the roof and the external margin of the orbit. It spares the frontal lobe since it does not necessitate cerebral retraction. This approach is useful for excision of accessible tumours from a frontal view by the superolateral and inferolateral quadrants, independently of their intra-conical or extra-conical location. As this approach permits access to the superior orbital fissure it concerns tumours situated, from a sagittal view in the two posterior thirds as defined by Benedict [4] (Fig. 11). Surgery should not include complex bone reconstruction after excision. In other words, it should be reserved for tumours of the soft intra-conical and extra-conical areas (for instance, neurilemomas, cavernous haemangiomas, dermoid cysts, etc.) according to the anatomical limits that we have identified and the biopsy or biopsy-excision with or without bone samples. The drawbacks of this approach are mini-

Fig. 10. The lateral intra-orbital approach (dissection): (a) The "S" shaped incision; (b) the periorbit is unfastened from the lateral wall of the orbit; (c) osteotomy takes the form of a dovetail; (d) the bone excision can be performed as far as the superior orbital fissure, towards the rear

mal provided the indication respects the anatomical limits that have been defined. From an aesthetic perspective, part of the incision is concealed in the eyelid (which must not be shaved); the posterior part of this latter bends towards the temple and can be usefully concealed in one of the wrinkles around the eyes. If the cutaneous closure is realised with care, it can become invisible within 3 to 6 months. The greatest obstacle is the frontal sinus. Before the age of ten years, it is little developed, if at all, and in adults can pneumatise the entire superior orbital margin as far as the external orbital process. Therefore, with this in mind, it is beneficial to use MR, and also CT, which provides more details of anatomical links with the frontal sinus. The frontal sinus may be opened accidentally or intentionally. If the opening is accidental, its size is generally small and the mucosa can be spared. There are more risks in ignoring the opening than there are difficulties in repairing it. In the case of accidental entry, the simplest method is to fill the opening with osseous powder and biological glue and close the gap from the exterior by applying a patch of temporal fascia stuck with acrylic-glue. When the frontal sinus has been opened intentionally it should be 'cranialised' by coagulating and resecting the mucosa of the sinus and filling the fronto-nasal canal with osseous powder and biological glue. The frontal sinus is the largest anatomical obstacle and brings many complications if these simple rules of exclusion are not respected.

The patient is in the supine position with their head turned 30 to 45 degrees towards the opposite side, in a simple headrest (a three pins holder cannot be safely used in children). The incision begins at the middle of the eyelid (it can be extended towards the interior, and only towards the interior if the objective is to gain access to roof of the orbit) and bends, in an 'S' shape towards the temple when the external orbital process meets the malar bone. At this stage, it runs alongside the superior rim of the zygomatic process (Fig. 10 a). The incision should go directly to the bone, and, if necessary the supra orbital nerve should be pushed aside from the supra orbital foramen. It is necessary to scrape the interior of the lateral orbital process and unfasten the periorbit from the lateral wall of the orbit (Fig. 10 b). The subcutaneous tissue is dissected from the fascia of the temporal muscle. Insertions of the temporal muscle are removed from the external temporal fossa and pulled towards the rear. This operation makes it possible to view the superior part of the external side of the great wing of the sphenoid. Osteotomy of the lateral orbital process may take the form of a step or, more simply, of a dovetail (Fig. 10 c). Once removed, the lateral wall of the orbit is gouged or drilled whilst keeping the periorbit at a distance. This bone excision can be performed as far as the superior orbital fissure, towards the rear (Fig. 10 d). Access to the intra-conical part of the orbit takes place between the superior and lateral rectus muscle. There is, therefore no interposition of important anatomical elements. By using this

approach it is possible to check the dura and remove the lateral part of the orbital roof by drilling the bone immediately below the line of insertion of the muscle, as far as is necessary in order to reveal the dura. This hole should be situated underneath the ridge of insertion of the muscle in order to conceal it during closure. This approach, when compared with a trans browsal approach, has the advantage of being able to remain strictly epidural, to check the orbital roof using both the intra orbital and epidural approaches if necessary. Finally, it is always possible to carry out a supra-orbital craniotomy by extending the cutaneous incision towards the interior of the eyebrow. However, this approach does not permit a satisfactory intra-dural opening or monitoring.

The Optic Nerve Approach

The optic nerve approach is justified for two kinds of tumours: meningioma of the optic nerve sheath and optic nerve glioma. In the first case, approaching the optic canal is the key point of the operation, and in the case of optic nerve glioma it is not absolutely necessary to open the canal although the optic nerve should be checked during its intra-dural course.

The Sub Frontal and Intra Conical Approach of the Orbit [7, 64]

The patient is in the supine position with their head towards the zenith or at a slight hyperextension of 10 degrees to distance the frontal lobe from the orbit, by gravity. After coronal incision the temporal muscle is pulled forwards en bloc with the periosteum and the skin flap. The notch or the supra orbital foramen is exposed with the supra orbital nerve. When it presents as a hole it should be open and the nerve individualized to be pulled forwards with the skin flap. A frontal craniotomy that is homolateral to the lesion is carried out. An accidental or intentional medial opening of the frontal sinus requires the same precautions to be taken as for a lateral approach of the orbit. In order to optimise the posterior field of vision and diminish cerebral retraction, it is possible to combine removal of the superior orbital margin after separating the supra orbital nerve from the orbital rim. It is possible, particularly with children, to carry out a combined approach in a single cranio-orbitotomy. The orbitotomy runs laterally as far as the lateral orbital process above the fronto-malar suture. It avoids opening the frontal sinus medially. The orbitotomy of the roof implies, in a first instance, perforating the roof with a one millimeter drill then detaching it like a postal stamp. The dura is separated from the orbital roof. This epidural separation does not, generally, allow the necessary space for exposing the whole of the orbital roof due to the impossibility of combining an epidural separation in the middle cerebral fossa. Under

these conditions it is advisable to approach the orbit by an intra-dural approach. Careful retraction of the frontal lobe can be easily obtained after opening the optico-carotid cistern and the slow, progressive subtraction of the Cerebro Spinal Fluid (CSF). Exposure of the two posterior thirds of the orbit towards the rear of the eyeball and across the entire width is then complete. The axis of the optic canal permits easy identification of the internal part of the orbital apex. If necessary, the superior orbital fissure can be opened after drilling the anterior clinoid process. A wide dural flap, attached by a pedicle to the median line, is pushed back medially to expose the orbital roof. By opening the optic canal it is possible to determine the apical area, which would be exposed across its entire width, and complementing surgical exposure using either a drill or a thin bone rongeur. The frontal nerve appears beyond the transparent periorbit: it is an excellent landmark before opening the periorbit, a longitudinal incision is made at the base of the levator palpebrae superioris muscle and completed by two perpendicular contra-incisions, one of which is anterior at the level of the eyeball and the other posterior at the level of the orbital apex. During this stage, any lesions to the trochlear nerve should be avoided as the nerve is situated medial to the frontal nerve, almost on the optic axis, in front of the roof of the optic canal. Orbital fat is usually considered as a dominating element of muscular function. However its coagulation and retraction appear indispensable in order to obtain a satisfactory view of the intra-conical anatomical elements. From the opened periorbit, there are 3 ways in which to penetrate the orbital cone.

The Lateral Approach is the Most Frequent (Fig. 12 a, b). It permits visualisation of lateral lesions to the optic nerve as well as those that spread from the superior orbital fissure to the lateral and apical area of the optic nerve. By pulling up the muscular levator palpebrae superioris/superior rectus muscles complex en bloc and using a siliconed surgical loop, it is possible to access the external part of the optic nerve. Lesions to the superior branch of division of the oculomotor nerve must be avoided whilst innervating the superior rectus muscle, setting the surgical loop sufficiently to the anterior, underneath the muscles. Pulled medially along with the other elements, the superior ophthalmic vein hinders the view of the optic nerve in its apical segment. It can be sacrificed or pushed laterally. The ophthalmic artery and the nasociliary nerve must be identified: they cross the superior side of the optic nerve, laterally to medially. At its medial bend the ophthalmic artery gives rise to the ciliar arteries and the lachrymal artery which is pushed away laterally. Further laterally the abducens nerve runs along the internal side of the lateral rectus muscle. The branch of division of the oculomotor nerve destined for the inferior oblique muscle is situated at the level of the inferolateral side of the optic nerve/ophthalmic artery

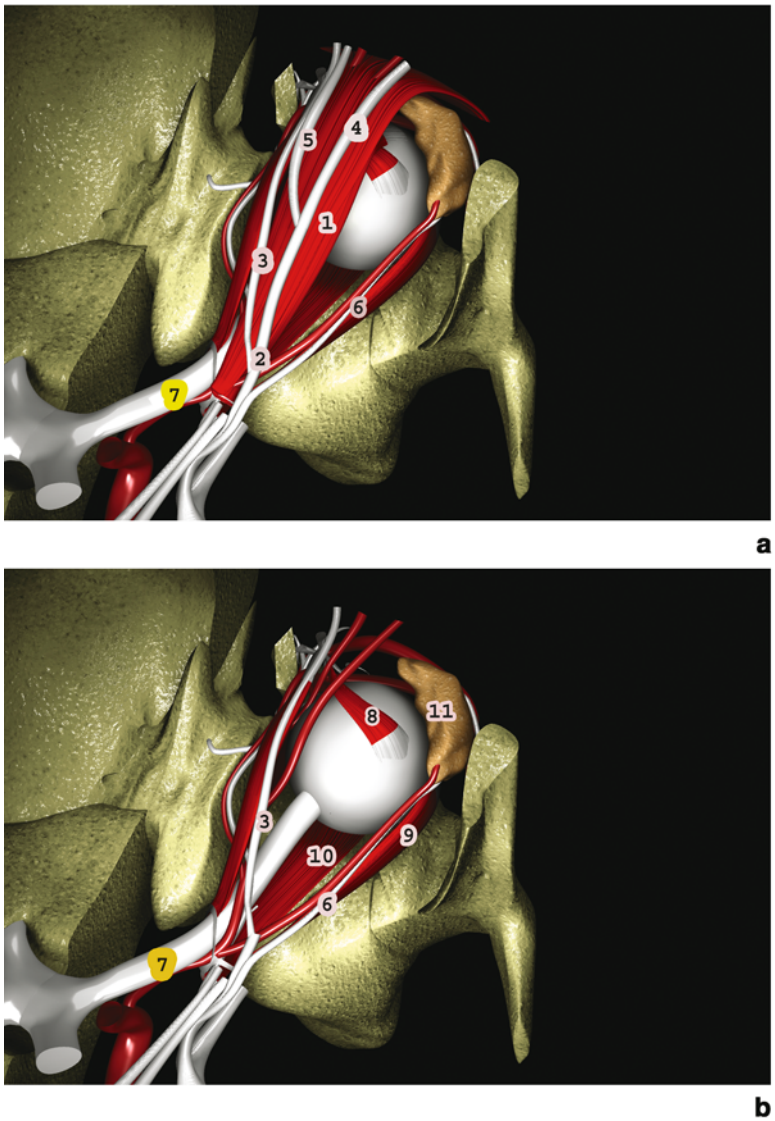


Fig. 12 (a–b): Superior aspect of the right orbit (unroofed) (a). Superior aspect of the orbital apex (The levator palpebrae and superior rectus muscles are reflected) (b). *1* Levator Muscle, *2* Frontal Nerve, *3* Trochlear Nerve, *4* Supra Orbital Nerve, *5* Supra Trochlear Nerve, *6* Lachrymal Nerve, *7* Optic Nerve, *8* Superior Oblique Muscle, *9* Lateral Rectus Muscle, *10* Inferior Rectus Muscle, *11* Lachrymal Gland

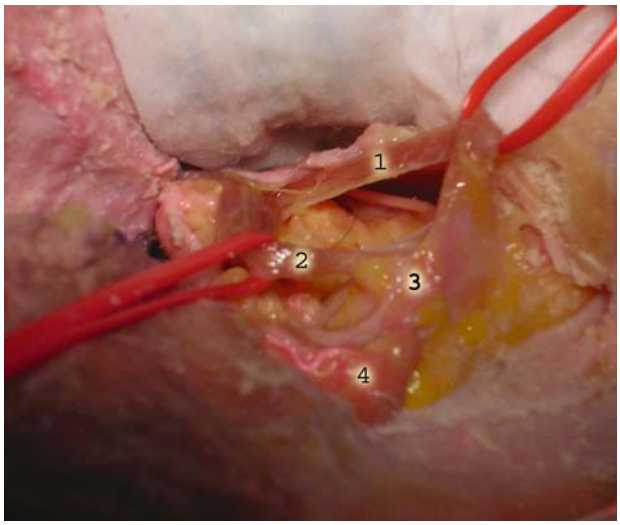


Fig. 13. The central intra orbital approach (dissection): *1* Levator Muscle, *2* Superior Rectus Muscle, *3* Whitnall's Ligament, *4* Lachrymal Gland

complex. The superior orbital fissure may be opened by making an incision to the annular tendon, backwards between the superior rectus muscle medially and the lateral rectus muscle laterally. By pushing apart the two muscular bodies at the level of the tendinous incision, the superior branches of division of the oculomotor nerve, the nasociliary nerve and the abducens nerve can be seen [20, 63, 64].

Central Approach (Fig. 13). This approach is most often indicated for limited surgery such as biopsy. As the levator palpebrae superioris muscle horizontally overlaps the medial side of the superior rectus muscle, it is possible to push the levator palpebrae muscle away medially and the superior rectus muscle laterally. Each muscle is retracted by a surgical loop. The frontal nerve can be kept in position and pushed medially with the levator palpebrae, or dissected then pushed away laterally with the superior rectus muscle. This permits a better view of the optic nerve at the level of the annular tendon. The trochlear nerve, at this level, is in an extra-conical position, directly in front of the optic nerve sheath and medial to the frontal nerve. It merits particularly careful consideration. This approach permits access to the entire width of the middle intra-orbital third of the optic nerve. However, it can be deleterious for the branch of division of the oculomotor nerve that innervates the levator palpebrae superioris muscle and should, therefore be undertaken with extreme care. Underneath this branch

of division that appears in the middle of the operating field of vision, the ophthalmic artery accompanied by the nasociliary nerve crosses the superior side of the optic nerve laterally to medially.

Medial Approach. This approach runs medially between the oblique superior muscle and the levator palpebrae superioris muscle and the superior rectus muscle laterally. The optic nerve is thus exposed across its entire length. The ophthalmic artery appears at the medial side of the optic nerve after crossing it. An incision may be made to the annular tendon towards the rear, between the levator palpebrae superioris muscle and the superior rectus muscle in such a way as to expose the optic nerve, at the level of the apex. During this incision, care must be taken to protect the trochlear nerve because it is in an extra-conical position.

Gliomas of the Optic Nerve

The slow or non-existent evolution of these tumours and the fact that vision is not affected make operations rare. The tumourous extension spreading backwards the chiasm is possible. The evolutive forms must therefore be operated on. After carrying out the sub frontal and intra-conical approach of the orbit the access to the intra-conical part of the orbit takes place between the superior and lateral rectus muscles. It is possible to access the optic nerve without removing the superior orbital margin and the adjacent part of the lateral orbital process. However, this orbitotomy increases the size of the area for using surgical instruments in the orbit and, to an equal degree, reduces the cerebral retraction which still remains epidural. An incision is made in the periorbit at the large axis of the orbit, parallel to the course of the optic nerve. The orbital fat will hinder access to the space between the lateral rectus muscle and the superior rectus muscle/levator palpebrae superioris complex. No dissection of the superior right muscle from the levator palpebrae superioris muscle must be performed as there is a risk of lesions to the nerves destined to the levator palpebrae muscle. It is necessary to retract the muscular group as a whole medially. The fat is the only remaining obstacle between the periorbit and the optic nerve. The tumour can simply be identified using one's finger before it is dissected and visualised. The fatty orbital body is pushed back medially with the superior muscle group and downwards with the lateral rectus muscle. The optic nerve is cut as closely to the surface as possible to the posterior pole of the eyeball. Dissecting the tumour is then realised backwards, still in the space between the superior muscular group and the external group, as far as the summit of the orbital cone where the optic nerve is cut behind the posterior pole of the tumour, which can then be removed. At this point there is a risk of wounding the ophthalmic artery medial or lateral to the

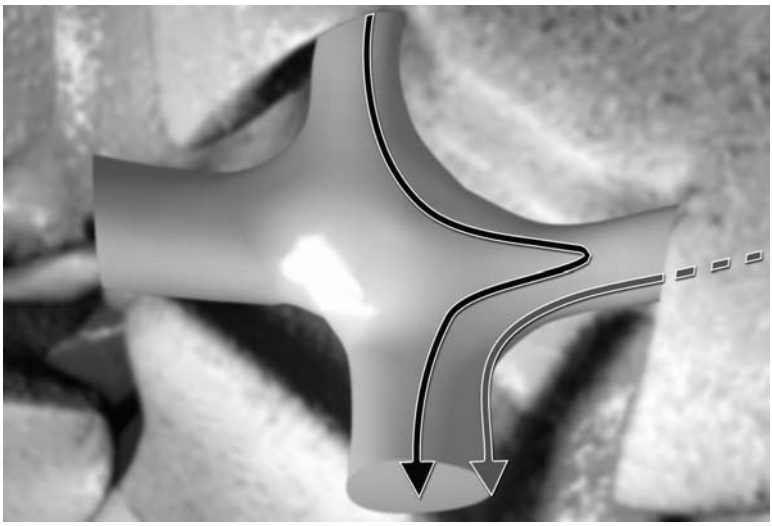


Fig. 14. Wilbrand's knee

optic nerve. This accidental injury is relatively harmless so long as the optic nerve is cut, but the difficulties of haemostasis in the fatty orbital body may lead to manoeuvres that are dangerous for the other nerve elements of the orbit. Furthermore, it is never certain that the incision to the optic nerve is in a safe area, since the incision is made as close to the outer cranial orifice of the optic canal as possible. Even though the tumour is strictly inside the orbit, it is preferable to complete the posterior incision of the intra-orbital optic nerve with a second intra-dural incision of the optic nerve, taking care not to open the optic canal. In this way it is possible to be sure that the second incision is made in a safe area. To do so, it is necessary to open the frontal dura. The frontal lobe must be carefully retracted half-way between a sub frontal and a pterional approach. The optic nerve is identified in the optochiasmatic cistern which is then opened. Resection of the optic nerve is carried out as far away as possible, forwards from the optic chiasm in order to avoid injuries to the nasal axons of the contra lateral optic nerve that have a short, recurrent passage in the optic nerve after crossing in the optic chiasm (Wilbrand's knee) [48, 59] (Fig. 14). The remaining part of the optic nerve in the optic canal is then simply co-agulated *in situ*. This second posterior incision protects the optic chiasm from a hypothetical posterior extension. If the tumourous extension directs itself backwards through the optic canal, it is approached in the same way as a meningioma of the sheath of the optic nerve, in order to resect the part of the tumour that lies in the optic canal Replacing the superior orbital margin and the part of the orbital roof which is adjacent to it, is generally

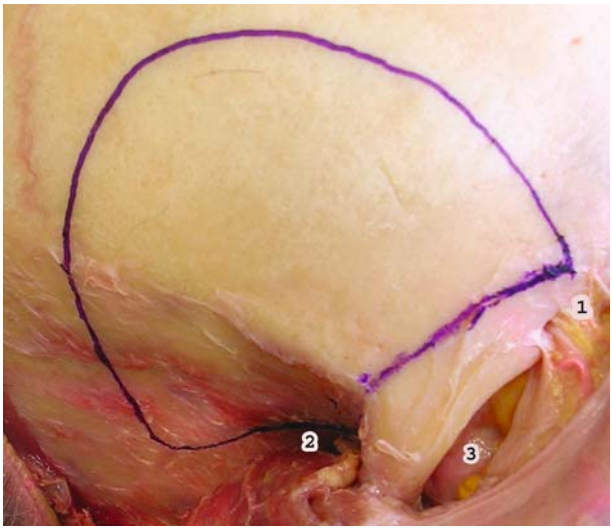


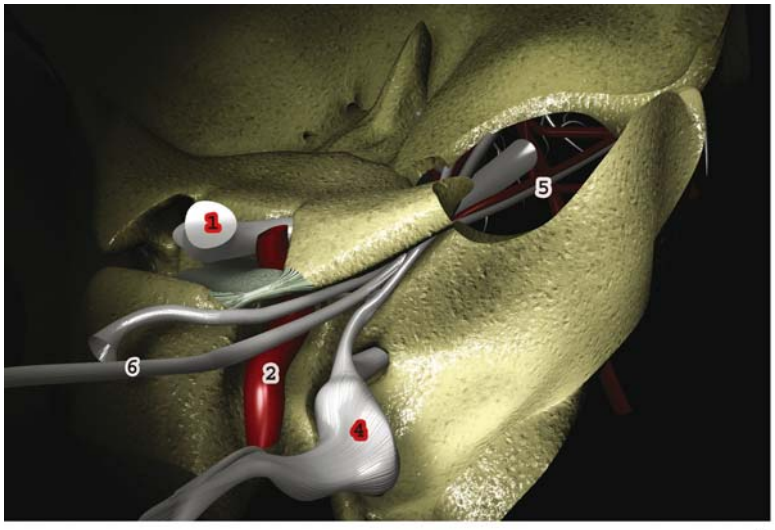
Fig. 15. Drawing of a fronto pterional craniotomy (dissection): *1* Supra Orbital Nerve, *2* Great Wing of the Sphenoid, *3* Lachrymal Gland

sufficient for reconstituting an osseous interface between the orbit and the dura of the inferior side of the frontal lobe, and does not necessitate a bone graft.

Frontopterional Approach of the Orbit

This surgical approach is mainly used for tumourous processes or pseudo-tumourous processes that are particularly extensive, like orbito-sphenoidal meningioma or fibrous dysplasia. It can be modulated according to the extension of the pathological process. The surgery is performed almost completely in an epidural situation after a wide separation of the dura above the orbit and of the temporal fossa [18]. The patient is in the supine position, the head in a slight hyper flexion to extend the field of operating vision at the level of the optic canal and the superior orbital fissure. The cutaneous incision is performed upwards from the tragus and follows, at an interval of 2 cm behind the hair implantation line until the frontal midline. The cutaneous point that corresponds to the pterion should be situated on line which runs from the two extremities of this incision. The skin, the pericranium and the temporal muscle are unfastened *en bloc* from the skull in order to avoid injuries to the frontal branch of the facial nerve. The fronto-pteriono-temporal area is thus exposed (Fig. 15). Some principles must be kept in mind in order to properly perform the craniotomy: it must be centred anteriorly by the pterion, and must be wide enough to

a



b

Fig. 16. Fronto ptérional approach of the right orbit (posterior and lateral aspects): (a) The pathological targets: the great wing of the sphenoid, the ridge of the lesser wing of the sphenoid, the superior orbital fissure; (b) the superior and lateral orbitotomy runs anteriorly to posteriorly from the pterion and the ridge of the lesser wing of the sphenoid; (c) the superior orbital fissure is opened; (d) the optic canal is opened and partially unroofed. *1* Optic Chiasm, *2* Intra Cavernous Carotid Artery, *3* Lesser Wing of the Sphenoid, *4* Trigeminal Ganglion, *5* Frontal Nerve, *6* Oculomotor Nerve

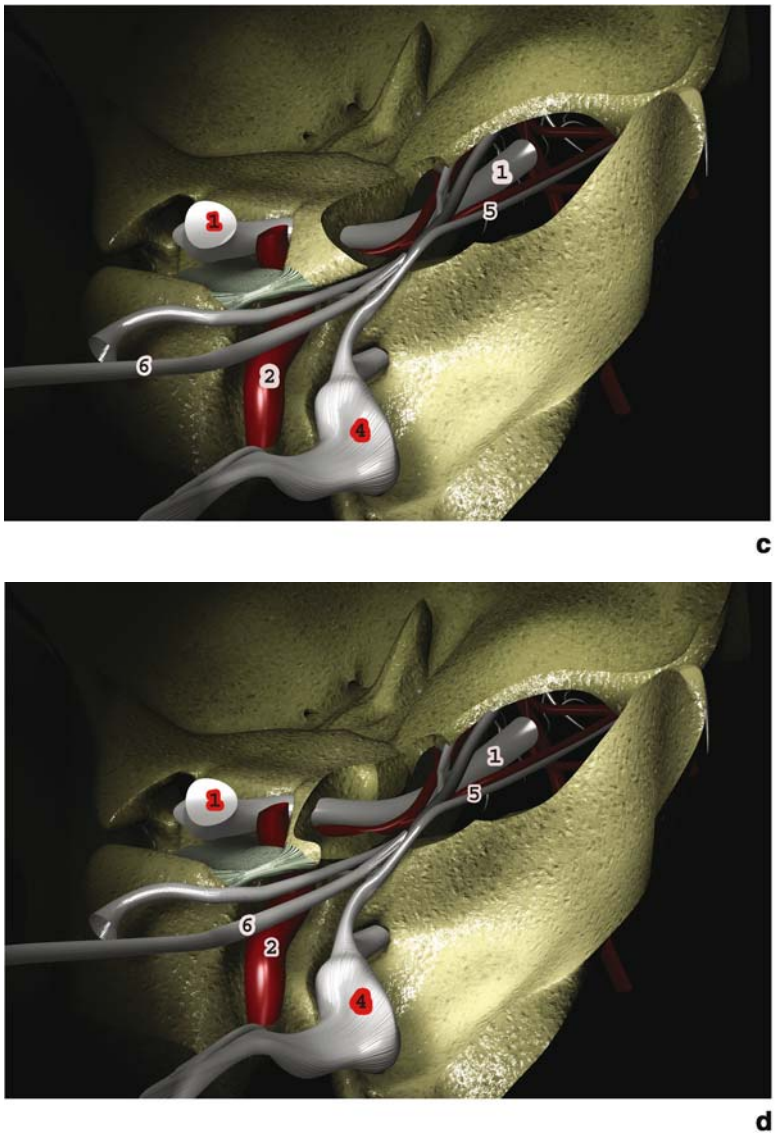


Fig. 16 (*continued*)

permit access to the pathological targets: the great wing of the sphenoid, the ridge of the lesser wing of the sphenoid, the superior orbital fissure (Fig. 16 a), the anterior clinoid process, the optic canal, the dura of the fronto-pterional-temporal area which is involved in the case of orbito-sphenoidal meningioma, opening the frontal sinus should be avoided unless it is particularly pneumatised and partially covers the orbital roof. This is a rare occurrence and the craniotomy in the frontal area has to follow an

angle of approximately thirty degrees from the horizontal plane. It is sufficient to expose the orbital roof without opening the frontal sinus. Multiplying the burr holes generally implies aesthetic sequelae. A single posterior burr hole in the hypothetic area the lateral fissure is often sufficient. This necessitates a slow and careful epidural unfastening which can be done with the Gigli's saw guide that permits an adapted length of interposition between the bone and the dura. Dural suspension should only be carried out at the end of the operation in order to avoid interfering with the epidural unfastening on the skull base. Removal of the superior orbital margin and of the zygomatic arch which is often advised [44] does not improve access to the posterior third of the orbit itself. This orbital-zygomatic removal can only be justified in the case of tumourous extensions towards the middle area of the temporal fossa, the petrous portion of the temporal bone and the pterygo-maxillar fossa. This technique does not strictly apply to the orbital approaches as we have defined them. The dura is opened then followed by the opening of the optico-chiasmatic and optico-carotid cisterns which allow gentle and progressive aspiration of the CSF thus reducing the brain volume in order to facilitate the epidural unfastening on the skull base. Often the dura is involved by the meningioma and we prefer to excise the pathologic area and repair it with an epicranial graft before drilling the skull base. The superior and lateral orbitotomy runs anteriorly to posteriorly from the pterion and the ridge of the lesser wing of the sphenoid (Fig. 16 b). It is necessary to begin drilling the superior and lateral wall of the posterior half of the orbit as the periorbit is more fragile at its anterior point. If the periorbit is torn, the gap will cause periorbital fat to escape, thus hindering the posterior field of vision for the surgeon. After having opened the superior orbital fissure (Fig. 16 c) and the optic canal (Fig. 16 d), the superior and lateral orbitotomy is completed forwards and terminates at the frontal and temporal bones. A large orbitotomy allows a partial reduction of the proptosis and the excision of the intra-orbital bud which spreads to the periorbit in an extra-conical position. The superior orbital fissure is opened after having drilled its roof; in other words, the ridge of the lesser wing of the sphenoid. This is the first orifice that is individualised. The anterior clinoid process lies posteriorly, the inferior and medial part of which is supported by the posterior root of the lesser wing of the sphenoid that corresponds to the lateral wall of the optic canal. Removing the clinoid process allows, on the one hand completion of excision when the pathological process concerns the anterior clinoid process and, on the other hand allows access to the optic canal via its external part [9]. Opening the optic canal is recommended as visual acuity will be impaired. The drilling at this point must be carried out using drills of 1 or 2 millimetres. Monitoring the optic nerve is carried out after opening the sheath in the optic canal.

Drilling the lateral part of the orbit terminates at the floor of the orbit, thus exposing the inferior optic fissure that links the inferior and lateral part of the orbit with the infra-temporal fossa on the one hand, and the pterygo-maxillary fossa on the other. Drilling of the temporal fossa is carried out downwards from the inferior orbital fissure and the foramen rotundum and oval posteriorly. It is important to avoid any contact with the periorbit as the orbit's fatty body will infringe onto the area of surgery, thus complicating the drilling operation considerably. If the periorbit is affected, it must be resected as much as possible. The periorbit is opened to the exterior of the frontal nerve that goes above and along the superior muscular complex. At the same level as the superior orbital fissure is the trochlea nerve which is situated medial to the frontal nerve, and is particularly vulnerable at this stage. The frontal nerve, which is a branch of the ophthalmic nerve, partially carries frontal palpebral sensitivity and is not a physiologically determining factor. The pathological process can spread backwards to the cavernous sinus. Its external wall can be peeled and coagulated without risk. A tumourous extension in the lateral wall of the cavernous sinus can be easily removed by excision, without risk. This is quite different to excision that is carried out in the cavernous sinus itself. Intracavernous surgery of meningeal residue is deleterious and this tumourous residue is easily accessible via radiosurgery or conformational radiotherapy. The periorbit is reconstructed using a fragment of pericranial graft sewn onto the periorbit which is considered to be safe. Bone reconstruction of the orbit is increasingly abandoned. As far as we are concerned, the only reconstructive surgery required concerns the defect of the frontotemporal bone defect (Fig. 1 c, d) which can be replaced by acrylic or a bone graft, thus compensating the atrophy of the temporal muscle and not allowing it to herniate through this bone defect which is often the cause of inaesthetic sequelae. The pathological limits are: periorbital, dural or osseous, whether radiological or macroscopic, and often very difficult to determine with precision; under such conditions the term of complete excision should be employed with great care. Intracranial neuronavigation offers a more precise schedule of excision when pre-surgical imaging and planning is carried out, but does not permit the application of a concept of excision that goes beyond extensions to these tumours, whose exact limits are not precise. However, only excision in safe osseous areas can minimise the possibilities of recurrences for the patient [49].

Problems of Orbital Reconstruction

Reconstruction of the orbit after a neurosurgical approach depends on the pathology concerned by the surgery, the extension of the osseous resections and on the nature of the bone which is sometimes affected by dysplasia.

The Orbital Rim

For aesthetic reasons it is always necessary to reconstruct of the orbital rim. Reconstructing the superior orbital and the lateral orbital rims is generally simple and necessitates either steel wire or screwed plates that may or may not be resorptive. This depends on each individual's preferences. In the case of bone defects concerning the orbital margin the same aesthetic reasons necessitate reconstruction. The most reliable method is an autologous graft. The most malleable substance and most suited to the constraints upon reformation of the orbital structure is the ribs, whether split or not. Harvesting is generally carried out before surgery. The number of sites of costal harvesting varies between 1 and 3.

The Orbital Walls (Fig. 8)

Small defects of the orbital roof do not require attention. However, large losses of substances necessitate reconstruction in order to maintain a solid interface between the dura of the frontal lobe and the periorbit. This reconstruction involves the split ribs when their removal has been made necessary in order to reconstruct the orbital structure or splitting the bone flap when removal of the ribs is not required. The bone graft is then simply inlaid or fixed from the superior orbital margin. It is not generally useful to reconstruct the lateral wall of the orbit. Even after complete resection of the tumour, the proptosis does not regress on occasions. This is why removing its external wall from the orbit plays such an important role in orbital decompression, as described in the guidelines for surgery for proptosis of Basedow's disease.

Orbital Dysplasia of the NF1

In the case of an orbital location of Recklinghausen's disease, it manifests itself as a plexiform neuroma and radically alters the social life of affected patients due to its appearance and the functional difficulty that it entails. The objective of therapy is to re-establish the balance between orbital content and container, thus requiring simultaneous surgery to the osseous structures (for example, reducing the increased size of the orbital volume) and the soft tissues (resection of the tumour). It is imperative, in the case of facial symmetrisation, that the volume of the bony orbit is reduced. Excision or iterative resections of tumours that are carried out on the soft tissues appear to be factors of less importance in the short and long term results. On a frontal view, it is necessary to restore the inferior and superior orbital margins, which are thinner than usual and irregular, whereas on a sagittal view is necessary, on the one hand, to reduce the hernia of the fron-

tal or temporal lobe and, on the other hand to set it by reconstructing the dehiscent orbital roof. The orbital roof, which is thinner and perhaps even absent, must also be reconstructed [28]. Indications for this depend on the size of the reduction in volume that is desired, and also on the preservation, or not, of visual function. Enucleation, despite allowing tri-dimensional control of the reconstruction operation, is always seen as a failure. Different approaches of reconstructive surgery have been suggested: the principle remains the same, which, in other words corresponds to a combination of separating the skull from the orbit, reducing the tumourous intra-orbital mass and remodelling the orbital skeleton. The osseous stage required for this operation associates to varying degrees, osteotomy and the on lay of organic material for grafts, or synthetic materials. Reconstruction of the orbit should begin with a transcranial approach [74], although the material to be used is still a subject of discussion [30]. Jackson [34] proceeds with the reduction of the hernia and contention of the temporal lobe by reconstructing the great wing of the sphenoid with the aid of split rib graft. Marchac [45, 46] proposes splitting the frontal bone flap to compensate for the bone dehiscence. Ulterior resorption of osseous grafts amongst this pathological tissue has caused certain authors to prefer this method to using acrylic [50] or titanium [74]. Reduction of the orbital content necessitates partial resection as total resection is rarely possible without inflicting serious functional impairment. Furthermore, in this instance the quality of resection depends entirely on the preservation of visual capacity. In many cases it is limited to tissular structures that are easily accessed by the superior defect. Remodelling the orbital skeleton aims to re-establish the balance between content and container by reducing the volume of the orbital whilst keeping the face symmetrical. This may necessitate osteotomy, but also appositional grafts or osseous substitutes. Associating osteotomy with autologous grafts would appear to yield better results in the long term. Jackson [34, 36] performs symmetrisation of the inferior orbital segment and the internal and lateral walls by means of osteotomy of the entire malar: this is possible by vertically cutting the internal wall of the orbit as far as the inferior orbital fissure. The zygomatic arch is sectioned and a horizontal maxillary osteotomy carried out below the infra-orbital nerve, in order complete the vertical osteotomy. Cutting the orbital floor facilitates the movement of the segment obtained in an upwards and medial direction. Any defect of maxillary bone can be compensated for with bone grafts, and the superior orbital margin benefits from split rib grafts. Munro [53] proposes inversed facial osteotomy in the event of maxillary asymmetry: this surgery combines a hemi-osteotomy of the type Lefort 1, and cranial-facial osteotomy of the type Lefort 3. The fragment that is obtained can be moved via posterior and superior rotation-translation. Facial symmetrisation is completed by bone grafts of iliac or costal origin Marchac [46, 47] completes an

osteotomy of the external orbital margin and repositions the canthal ligaments. He combines this with the installation of osseous grafts to the orbital roof. Remodelling of the soft tissues follows osteotomy because, if the eyelid is affected this contributes to the imbalance between content and container. As far as possible, surgery at this stage should respect its function. This requires transfixing resection of the eyelid on horizontal and vertical planes, whilst taking care to maintain the functioning of the levator palpebrae superioris muscle. Marchac [45, 47] carries out resection of the cutaneous excedent, on the whole thickness by comparing the unaffected side. The levator palpebrae superioris muscle is re-inserted after reduction of its movement on the tarsus. An external canthopexy would conclude surgery. This pathology demands delicate surgery both to the osseous structures but also to the soft tissues. This is all the more true as surgery may still engender post-surgical complications [62]. However, it does offer aesthetic and functional solutions that may be partial and imperfect, but are desired by patients.

Surgery in the Case of Fibrous Dysplasia (FD)

Modelling and conserving resection in fibrous dysplasia reduces the undesirable aesthetic element, but does not control the potential growth of the remaining process and the recurrence of deformation. Total or partial resection of the pathological tissue followed by reconstruction during the same operating stage would seem preferable [54, 17]. This method is based on ablation of the entire pathological process by an approach that may be intracranial when the FD is at the base of the skull, or facial when the FD has a lower extension. The surgical approach is endo cranial, facial or combined according to the size of the FD extension at the base of the skull. Reconstruction necessitates autologous bone grafts [17, 24, 35, 53, 54, 77] proposes reinstating the pathological tissue after remodelling. Decompression of the optic nerve in the optic canal follows the same principles as for the epidural approach of the optic canal in the case of orbito-sphenoidal meningioma. The indication of decompressing the optic canal is given when there is a reduction in visual acuity due to the optic canal being affected by the FD or when surgery is justified on the grounds of aesthetic consideration and there is a threat of compression of the optic nerve by its surrounding bone structures [58]. The epidural and intra-orbital endo cranial approach yields the best results as it allows decompression of the whole course of the optic nerve. Moreover, accurate identification of the different anatomical elements makes this surgical approach safer and more secure [13]. The main difficulty in this type of surgery is linked to the thickness of the dysplastic bone since the roof of the orbit can, on occasions reach a thickness of 1 cm.

Conclusions

Neurosurgical approach of orbital tumours concern the posterior two thirds of the orbit. Whether concerning intra- or extra-conical pathologies, it is their direct or indirect repercussion on the optic nerve that, essentially justifies the neurosurgical indication. Any lesion that necessitates an intra-operative access to of the orbit, its contents and its peri-cerebral spaces should be approached by using one of the neurosurgical techniques that we have discussed. The current methods of diagnosis by imaging should make it possible to foresee the degree, and succession of surgery required. As a result of these findings, a pluridisciplinary approach is organized and the specific therapeutic orientations of each tumour can be considered.

Acknowledgements

We thank Professor Marc Braun of the University of Nancy (France) for allowing realisation of the dissections displayed in this chapter.

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