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SHORT COMMUNICATION

A sphenoid en plaque meningioma aggravates exophthalmos in a patient with thyroid ophthalmopathy

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PURPOSE. *To describe the rare case of a patient with thyroid ophthalmopathy whose unilateral aggravated exophthalmos was caused by the development of an ipsilateral sphenoid wing en plaque meningioma.*
METHODS. *Case report.*
RESULTS. *The ophthalmologic examination included visual acuity assessment, anterior segment examination, funduscopy, ocular motility examination, and exophthalmometry. Magnetic resonance imaging (MRI) of the brain and orbit revealed the existence of a sphenoid meningioma. Removal of the tumor through a pterional craniotomy was performed. The histologic examination showed that it was an en plaque meningioma. There were no postoperative complications and no recurrence of the tumor was revealed 24 months after the operation.*
CONCLUSIONS. *Detailed ophthalmologic examination and MRI of the brain and orbit are necessary in every patient with deterioration of the exophthalmos even if the suggestive cause seems obvious. (Eur J Ophthalmol 2006; 16: 461-4)*

KEY WORDS. *En plaque meningioma, Thyroid ophthalmopathy, Orbital tumor*

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INTRODUCTION

Meningiomas are benign slow-growing tumors that arise from arachnoid cap cells. They are the second most common brain tumor in the adult population. Female:male ratio is 1.8:1 and the usual age at occurrence is between 30 and 50 years. In the orbit, they are observed four times more frequently than astrocytomas (1). Meningiomas are the most common tumors that may develop following radiation treatment of the central nervous system or the surrounding tissues (2). The symptoms and clinical signs of meningiomas depend on the location of the tumor. The most important types of meningiomas that are located in the orbit are 1) the primary meningiomas of the optic nerve and 2) the sphenoid wing meningiomas. Early diagnosis is important because total removal of the tumor may be feasible and the vision of the patient can be preserved.

En plaque meningiomas are rare tumors, which are usually located in the sphenoid wing and rarely in other locations. These tumors are likely to provoke hyperostosis, which is often disproportionate to their relatively small size. It is these bony changes that produce the clinical symptoms and signs (3). En plaque meningiomas invade the brain and the orbit through natural anatomic foramina and fissures, as well as through penetrating vessels that are abundant in the area around the tumor (4).

The aim of this study is to describe a patient with thyroid ophthalmopathy, in whom aggravation of already existing exophthalmos was due to the development of a sphenoid wing en plaque meningioma and not to the existent disease.

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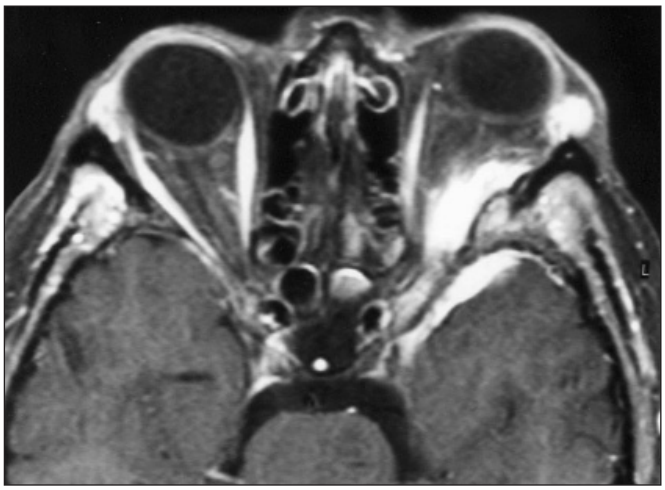

Fig. 1 - *Preoperative magnetic resonance imaging of the orbit and brain with gadolinium, showing the en plaque meningioma extending to the orbit.*

Case report

A 61-year-old woman was admitted to the Department of Ophthalmology with visual disturbances of the left eye for 2 months prior to her admission, and progressive deterioration of a preexisting exophthalmos of the same eye. She had a history of hyperthyroidism under treatment for 8 years. Her past ophthalmologic history was significant for dysthyroid ophthalmopathy with bilateral exophthalmos starting 4 years prior to her admission.

She was never submitted to any kind of radiation therapy. Her visual acuity was 20/25 in the right eye with –0.75 sph and 20/30 in the left eye with –0.5 sph -0.5 cyl x 90°. There was hyperemia of the conjunctiva, slight lid retraction, and nuclear cataract in both eyes. The fundus was normal and the intraocular pressure was 15 mmHg in the right eye and 16 mmHg in the left. The exophthalmos (measured by the Hertel exophthalmometer) was 22 mm in the right eye and 25 mm in the left eye (base 110 mm). Extraocular motility was full. Pupillary reactions were normal. Goldmann perimetry demonstrated defect in I_3 and I_2 isopters, more intense in the left eye. MRI of the brain and orbit revealed a homogeneously enhancing tumor extending from the cavernous sinus along the dura of the ipsilateral (left) temporal and anterior cranial fossa.

The lesion expanded through the left optic canal to the left orbit. Hyperostosis of the ipsilateral sphenoid wing was also observed. All these findings were suggestive of a meningioma (Fig. 1). Due to the fact that hormones and especially progesterone influence the tumor growth in

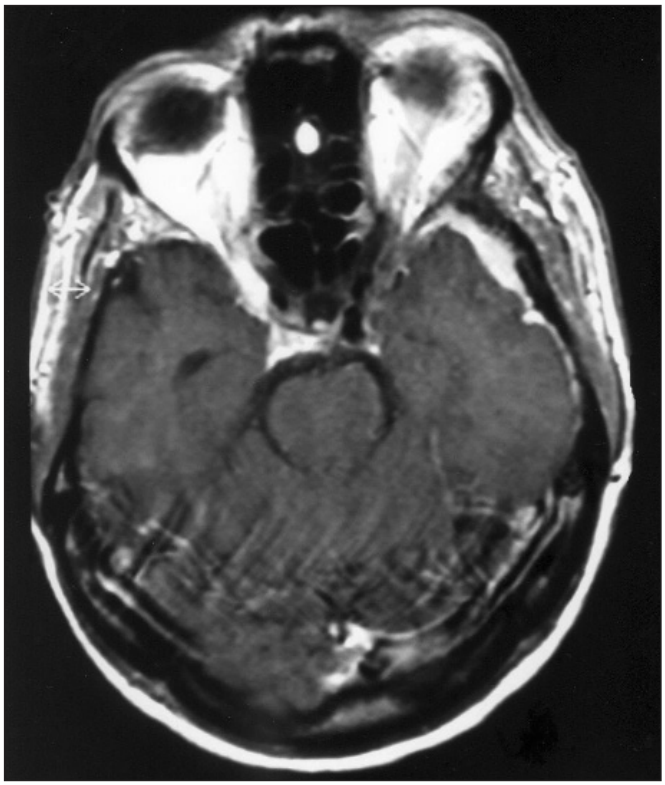

Fig. 2 - *Postoperative magnetic resonance imaging of the orbit and brain with gadolinium 2 years after the operation. No recurrence of the en plaque meningioma is observed. The intraorbital part of the tumor remains unchanged.*

meningiomas, our patient underwent a detailed clinical and laboratory examination by an endocrinologist that revealed no abnormal findings (4). Through a pterional craniotomy the intracranial part of the tumor was removed. The postoperative course of the patient was uneventful. The histologic examination showed a meningothelial growth type meningioma. The postoperative MRI, 2 years after the operation, did not reveal any tumor recurrence (Fig. 2). Twenty-four months after the operation, the patient had visual acuity 20/25 in the right eye, 20/25 in the left, with correction. There was pseudophakia in both eyes due to cataract surgery. Fundus, extraocular motility, and intraocular pressure were normal. Exophthalmos and visual fields remained unchanged.

DISCUSSION

En plaque meningiomas are relatively rare tumors. Except from isolated cases, a few series are found in the literature. They are almost exclusively found in females

(young or middle aged) and located in the sphenoid bone (5). Previous case reports have shown the association of these tumors with visual and hearing impairments and also with malignancy and metastases (6, 7).

The main symptoms leading the patient to an ophthalmologist are as follows, in decreasing frequency: proptosis, vision impairment of the ipsilateral eye, retroorbital pain and lid edema, visual field deficits, and diplopia. These are caused by the chronic pressure on the optic nerve and the difficulty of movement of the eyeball inside the orbit (3, 4). Visual loss due to pressure on the optic nerve and subsequent atrophy are rare. Papilledema may sometimes be observed and is related to the pressure on the optic nerve by the tumor and the surrounding edema and not to the increased intracranial pressure. Our patient had normal fundus examination on both eyes. Preoperative fields examination revealed a defect in I_2 and I_3 isopters.

Meningiomas are frequently accompanied by skull abnormalities. Hyperostosis is much more common than bone destruction by the tumor. In the case of en plaque meningiomas the tumor cells are located in the haversian canals. The extent of bone destruction will determine the type of operation and the probability of tumor recurrence (4). Computed tomography (CT) and MRI of the brain and orbit are the neuroradiologic diagnostic methods of choice (4, 8). CT shows only the hyperostosis, while MRI shows the soft part of the tumor, its relation to the brain and nerves, and meningeal involvement. When there is extension of the tumor inside the orbit, as in our case, CT and MRI can reveal the intraorbital part of the tumor, as well as the optic nerve encroachment.

Differential diagnosis of the en plaque meningiomas must be made from other space occupying lesions as fibrous dysplasia, osteoma, Paget's disease, hyperostosis frontalis interna, sarcoidosis, and the primary intraosseous meningioma (9-11). En plaque meningiomas are characterized by a great degree of infiltration of the dura. In cases with a lesser degree of dural infiltration, the differential diagnosis from the primary intraosseous meningioma may be very difficult.

In the past, the results of the surgical treatment of en plaque meningiomas were disappointing but today, radical removal seems necessary and generally accepted. First of all, the patient is relieved from all known consequences, such as tumor extension, dura infiltration, and bone destruction. Secondly, viability of the optic nerve and visual function are assured since there are no compressive effects inside the orbit (12). The postoperative visual function of our patient was good, both in the immediate postoperative period and in the last follow-up, 2 years later.

Regarding the postoperative exophthalmos, no major difference was observed. The difference of 3 mm remains stable 2 years after the operation. No diplopia was observed in the postoperative period, as well. Honeybul et al, postoperatively, report no change in visual acuity in 12 of their 15 patients, no permanent diplopia in 13 of them, and improvement of the exophthalmos in 12 of them (3).

En plaque meningiomas that extend in and around the orbit do recur (3). The main cause of recurrence is the limited resection of the infiltrated bone and dura (13). Total removal is usually achieved for tumors in external and middle-third locations, but it is more problematic in tumors extending along the whole sphenoid ridge or located at the inner third. Subtotal extensive removal, however, combined with decompression of the cranial nerves and orbit, usually allows good functional and cosmetic results (5). Proptosis or deterioration of postoperative exophthalmos are the main clinical signs of recurrence. In our patient, both clinical evaluation and neuroradiologic examinations 24 months after the operation did not reveal any tumor recurrence.

Our case is noteworthy because our patient had an aggravated exophthalmos for a couple of months, which could be easily attributed to the already existing thyroid ophthalmopathy. However, the occurrence of an en plaque meningioma with extension to the left orbit turned out to be the cause of this exophthalmos. According to our knowledge, this rare coexistence of these two entities has not yet been described and it is observed only in cases with previous radiation treatment for Graves' disease (2). Therefore, clinicians should be alerted from any change of the clinical signs and symptoms, especially if they are unilateral and unrelated to the outcome of the underlying disease. This case also emphasizes the value of CT and MRI in the follow-up of patients with thyroid ophthalmopathy.

The authors have no proprietary interest in this study.

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