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SHORT COMMUNICATION

Sarcomatoid carcinoma and orbital apex syndrome

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PURPOSE. *To report a case of sarcomatoid carcinoma and orbital apex syndrome in a previously healthy adult.*

METHODS. *A previously healthy 45-year-old man presented with exophthalmos of the left eye and a mass visible through his left nostril. A biopsy was performed and immunohistochemistry was used to confirm the diagnosis of the tumor. The patient was treated with chemotherapy and radiotherapy.*

RESULTS. *After treatment, we observed a substantial reduction in the size of the mass, but side effects of treatment developed. The visual acuity of the left eye was no light perception. Eight months later, the patient presented with bone and liver metastases, and he died 4 months later.*

CONCLUSIONS. *Sarcomatoid carcinoma is an aggressive tumor that can produce compressive symptoms with very poor visual and survival prognoses. A cranio-orbital computed tomography scan should be performed when patients present with symptoms of conjunctivitis and orbital apex syndrome. (Eur J Ophthalmol 2006; 16: 608-10)*

KEY WORDS. *Optic disc atrophy, Orbital apex syndrome, Sarcomatoid carcinoma*

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INTRODUCTION

Sarcomatoid carcinoma is a rare, aggressive tumor, with a special predilection for the upper bronchodigestive tract and oral cavity (1-3), but it may occur in other locations such as the uterus and breast. This tumor has a mixture of carcinoma and sarcoma containing differentiated mesenchymal elements (1-3).

Orbital apex syndrome is a rare complex characterized by proptosis and paralysis of extraocular muscles, associated with involvement of the first division of the trigeminal nerve and visual loss (4). The most common diseases responsible for this syndrome can be metastatic tumors, orbital cellulitis (bacterial infection), orbital myositis, and fungal infection (4). To our knowledge, this is the first case of sarcomatoid carcinoma of the left ethmoid sinus presenting as orbital apex syndrome.

Histologic aspects of this tumor are areas with spindles and epithelial characteristics cells. The tumor cells express vimentin and cytokeratin staining. Immunohistochemistry was used to confirm the diagnosis.

Case report

A 45-year-old man was referred to the ophthalmology clinic with a 10-month history of eyelid swelling and bilateral conjunctivitis. Physical examination showed pain, diplopia, and axial proptosis of the left eye, and a mass was visible through his left nostril. The visual acuity was 20/20 bilaterally. External examination of the left eye showed periorbital swelling, ptosis, and chemosis (Fig. 1). There was no afferent pupillary defect, no impairment of color vision, and no corneal sensation. The left eye had a blurred optic disc, choroidal folds, and vascular congestion (Fig. 1); the right

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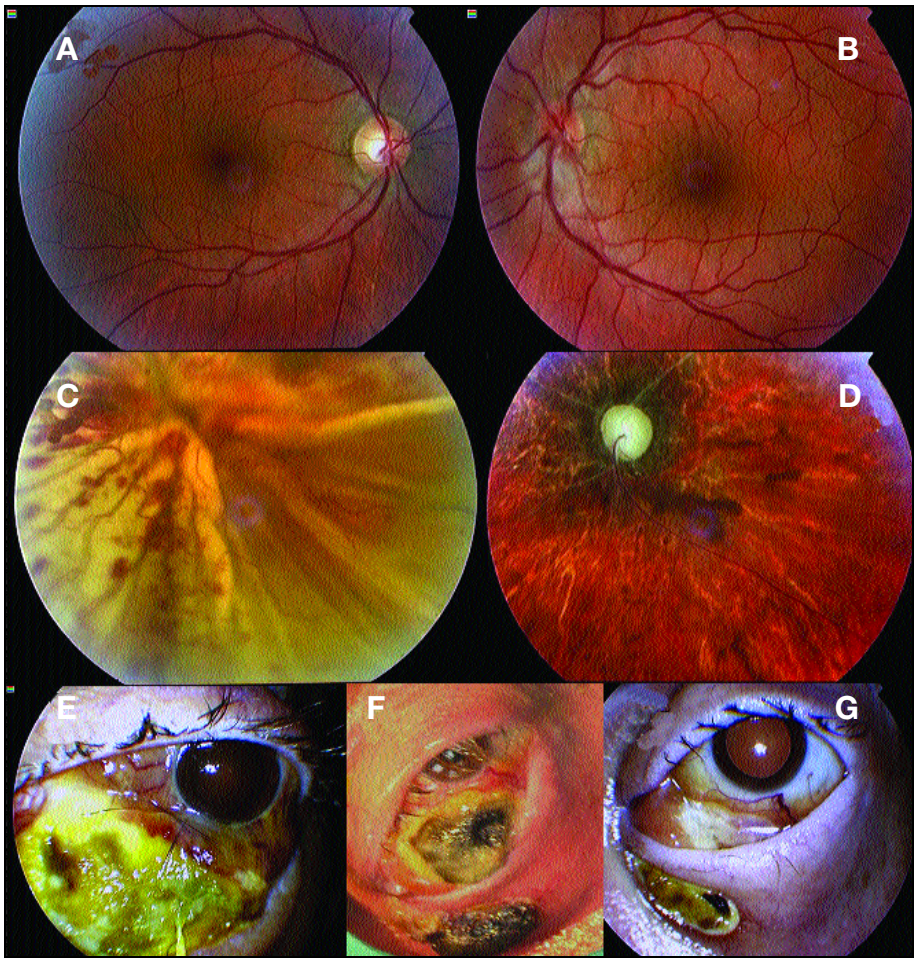

Fig. 1 - *(**A**) No abnormalities are seen in a fundus photograph of the right eye. (**B**) The left eye has a blurred optic disc, choroidal folds, and vascular congestion. (**C**) Two months after treatment, obstruction of the arterial and vein of the retina, hemorrhages, and exudative retinal detachment are seen in the left eye. (**D**) Four months later, optic nerve atrophy is seen, but no retinal detachment. (**E**) External photograph of the left eye shows periorbital swelling, ptosis, and chemosis. (**F**) External photograph of the left eye 2 months after treatment. (**G**). External photograph of the left eye 4 months after treatment.*

eye was normal (Fig. 1). A cranio-orbital computed tomography (CT) scan (Fig. 2) showed a large mass arising from the maxillary sinus that eroded the floor of the left orbit involving the left retrobulbar space and extended to the apex of the orbit to the optic chiasm.

The mass was biopsied. Five-micron sections were cut and stained with hematoxylin-eosin (Fig. 2). Immunohistochemical study showed positive staining with vimentin for the epithelial markers cytokeratin (CAM 5, CAM 2), vimentin (Fig. 2), and S100 protein. A diagnosis of sarcomatoid carcinoma was made (2, 5) and no distant metastatic lesions were found. Because the tumor was considered too extensive for surgery, the patient was treated with external radiotherapy (64.5 Gy) and chemotherapy (paclitaxel, 5-fluouracil, and cisplatin). Two months later, we observed a reduction of the mass (Fig. 2), and the external examination of the left eye also improved (Fig. 1). The visual acuity of the left eye was no light perception and the funduscopic examination (Fig. 1) showed obstruction of the retinal artery and veins, hemorrhages, and an exudative retinal detachment. Four months later, we observed optic nerve atrophy and the vision was no light perception. Eight months later, the patient presented with bone and liver metastases and died 4 months later.

DISCUSSION

The differential diagnosis of orbital apex syndrome is extensive, and neither the mode of onset nor the evolution of neurologic deficits can distinguish among a neoplasm, an aneurysm, or inflammatory processes (5). An orbital infection, either bacterial or fungal, is consistent with this presentation, but this patient was not immunosuppressed. Any inflammatory process, such as Wegener's granulomatosis, sarcoidosis, Tolosa-Hunt syndrome, or orbital inflammatory pseudotumor, can cause a rapidly

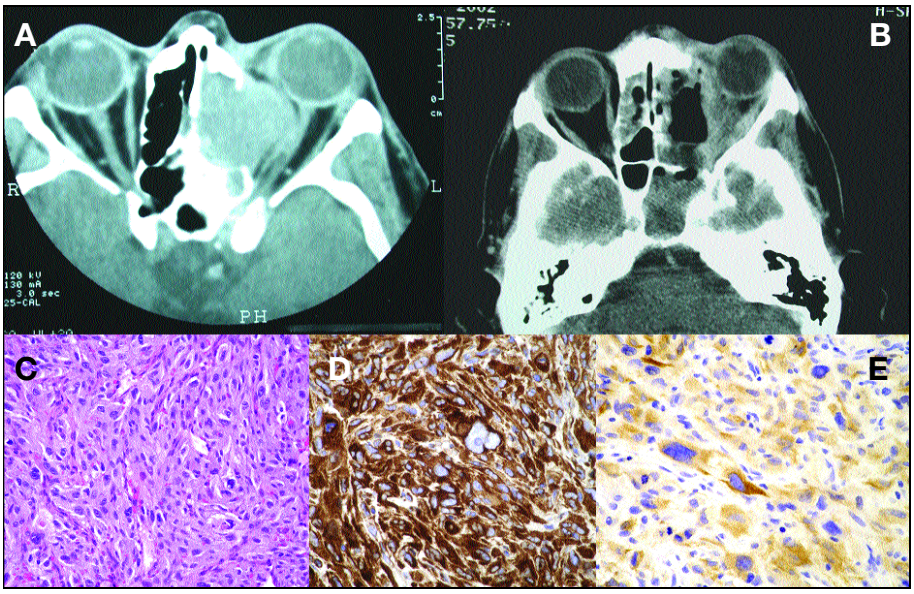

Fig. 2 - *(**A**) Cranio-orbital computed tomography scan shows a large mass arising from the maxillary sinus, eroding the floor of the left orbit involving the left retrobulbar space, and extending to the apex of the orbit to the chiasm. (**B**) Two months later, a dramatic reduction of the mass is observed. (**C**) Areas with spindles and epithelial characteristics cells are observed (hematoxylin-eosin x400). (**D**) Vimentin staining. The tumor cells express vimentin (immunohistochemistry, original magnification x100), especially staining of spindle cells. (**E**) Areas of spindle cells are focally and weakly positive for cytokeratin staining (original magnification x100).*

evolving orbital apex syndrome (4). CT scan and the biopsy established a definitive diagnosis.

The cause of the optic nerve atrophy and the retinal vessels obstruction was the sudden growth of the tumor. Although radiotherapy and chemotherapy reduced the mass it was too late and optic nerve atrophy was definitive.

This case is unique because the patient had had conjunctivitis for approximately 1 year, and the symptoms developed only when the tumor enlarged. A cranio-orbital CT scan should be performed immediately when patients present with symptoms of orbital apex syndrome. It is important to make the correct diagnosis because once treatment is started favorable visual and survival prognoses might be achieved.

None of the authors has a financial, commercial, or proprietary interest in any of the instruments or intraocular lens mentioned in this report.

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