
SHORT COMMUNICATION

Case report

Intraocular invasion by malignant orbital fibrous histiocytoma: a case report

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PURPOSE. *To present a case of intraocular invasion by an orbital malignant fibrous histiocytoma.*

METHODS. *Case report.*

CASE REPORT. *An 84-year-old man was exenterated due to a malignant fibrous histiocytoma of the right orbit. On image investigation and histologic sections, the tumor entirely filled the right eye and orbit. The patient died of intracranial spread of the lesion.*

CONCLUSIONS. *Fibrous histiocytoma of the orbit can invade the eye. (Eur J Ophthalmol 2001; 11: 306-8)*

KEY WORDS. *Malignant fibrous histiocytoma, Intraocular invasion, Orbital tumor*

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INTRODUCTION

Fibrous histiocytoma is one of a group of tumors of mesenchymal origin and varied clinical behavior. The Armed Forces Institute of Pathology reports that it is the most frequent mesenchymal tumor arising in the orbit (1) but a 40-year study at the Mayo Clinic found fibrous tumors were responsible for only 1.5% of 1376 cases (2). This wide variation is best explained as a referral bias.

These tumors are most frequently localized in the medial and superior orbit, but can also be found in the conjunctiva and lacrimal drainage system (1, 3, 4). They are reliably classified histologically as benign, intermediate or locally invasive, and malignant forms. The clinical evolution corresponds well with the histology. The malignant form is the most rare (1).

This case report describes a malignant orbital fibrous histiocytoma with intraocular invasion.

Case report

An 84-year-old man presented to the Ophthalmological Service of the Federal University of Bahia,

Brazil, with a history of a right orbital lump for 8 months. He reported loss of vision in that eye following blunt trauma 15 years ago. Ocular examination found a hard orbital mass protruding through the right lid aperture. The mass was covered by hyperemic conjunctiva. No pre-auricular or sub-mandibular nodes were palpable. The left eye had visual acuity 20/25 and no other abnormality.

Computerized tomography revealed a soft tissue mass that filled the right orbit including the globe. Some areas of hypodensity were suggestive of necrosis. The bony walls were thinned but intact, without invasion of the sinuses. Biopsy was carried out, establishing a diagnosis of malignant fibrous histiocytoma. A partial exenteration was done, sparing lid skin.

Four months after surgery, the patient returned with the right orbital cavity partially filled by a violet mass and a right pre-auricular node measuring 2 x 2 cm. The patient was referred to a clinical oncology service for treatment. He developed intracranial spread of the lesion and died in spite of chemotherapy and radiotherapy.

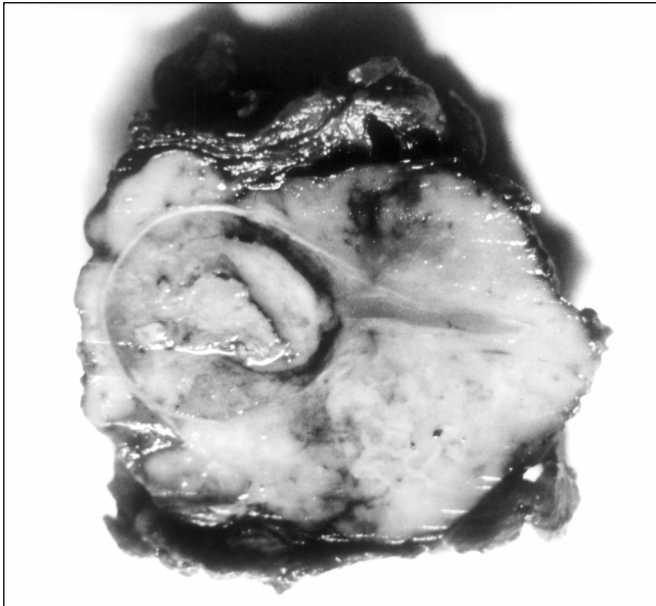


Fig. 1 - Tumor mass filling the left orbit and replacing intraocular contents through a posterior scleral defect.

Macroscopy

The right orbital contents measured 54 x 50 x 49 mm. The cut surface of the tumor was white and yellow. The intraocular structures were completely replaced by the tumor that had apparently entered the globe through a wide posterior scleral defect (Fig. 1).

Microscopy

The tumor was composed of spindle cells arranged in a storiform pattern and histiocytoid cells with clear and foamy cytoplasm (Fig. 2). Nuclear pleomorphism, multinucleated giant cells and some areas of coagulation necrosis were present (Fig. 3). A delicate fibrillar meshwork was seen on Picrosirius Red staining. The foamy cells stained positively for anti-CD 68 antibody and negatively for anti-desmin antibody.

DISCUSSION

The malignant variant of fibrous histiocytoma is the most rare of all, according to Font and Hidayat in the largest series published of 150 cases from the Armed Forces Institute of Pathology, being responsible for

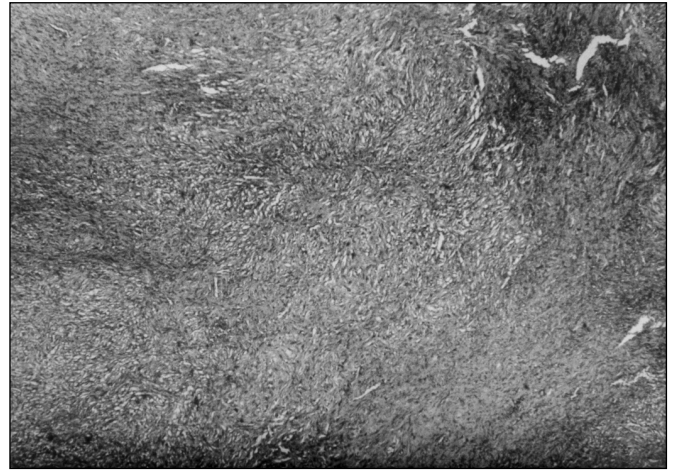


Fig. 2 - Spindle cells in a storiform pattern and histiocytoid foamy cells (Hematoxylin-Eosin 100x).

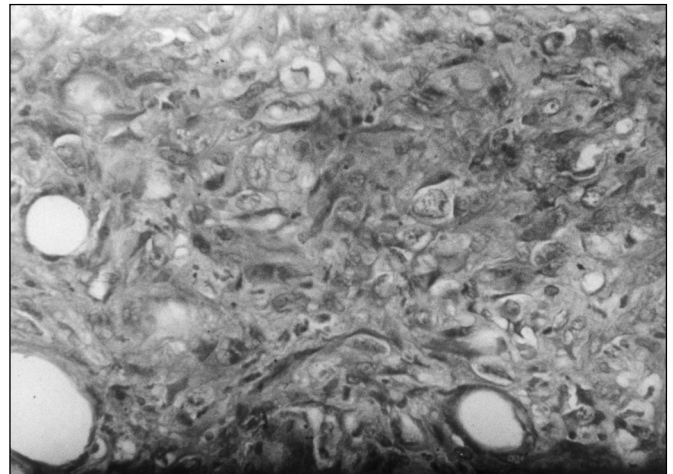


Fig. 3 - Anaplastic and multinucleated giant cells (Hematoxylin-Eosin 400x).

11.4% of cases (1). The clinical picture is usually of rapid onset (mean duration 3.4 months) with proptosis, orbital mass and decreased vision the most common findings (1). Computerized tomography and magnetic resonance imaging are of value to outline the tumor and facilitate a precise surgical approach, but of less value in the final diagnosis (5). The treatment of choice is complete surgical excision, which is difficult since the tumor is not encapsulated and has infiltrating edges (1, 2). Adjuvant treatment includes radiotherapy and chemotherapy, but neither seems to have much effect. In contrast to the benign and locally invasive variants, the malignant form has

a very poor prognosis with a ten-year survival rate of 23%. Local recurrence can be as high as 64%. The tumor can cause death by direct invasion of adjacent structures or lymphatic and hematogenous metastases. These have been reported to appear even two decades after an apparently successful excision (1).

The histological criteria to classify a fibrous histiocytoma as malignant include nuclear pleomorphism, multinucleated giant tumoral cells, areas of necrosis and mitotic activity (1). The clinical and histopathological diagnosis is difficult. Differential diagnosis includes hemangiopericytoma, hemangioendothelioma, angiosarcoma and inflammatory granuloma (1, 6). The exact diagnosis can be confirmed immunohistochemically or by electron microscopy (3, 6). Intraocular invasion by an adnexal tumor is a rare and usually late event. It is more commonly related to epithelial tumors of the ocular surface, which have either been neglected or incompletely excised (7). Orbital tumors tend to indent and dislodge the globe, rather than invade it (2).

Despite the invasive potential of malignant fibrous histiocytoma, no report of intraocular spread could

be found in the literature. In our case, the history of ocular blunt trauma may have caused a posterior scleral rupture or weakness that facilitated the intraocular spread of the lesion, since this is not its characteristic behavior.

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