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Case Report

Epithelioid hemangioendothelioma, an uncommon tumor of the eyelid: A case report

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ABSTRACT

Epithelioid hemangioendothelioma (EHE) is an infrequent vascular tumor that has an intermediate prognosis between the prognosis of the angioma and that of the angiosarcoma. Its evolution is characterized by a tendency to recurrence and metastases. EHE has been reported in locations such as: lungs, liver, bone and soft tissues. There are very few reports of cases involving the head and neck region. The occurrence of EHE in the orbital region and/or the eyelid is extremely rare. The authors report a case of EHE in the right upper eyelid. Surgical excision has been performed resulting in a good local control of the disease after a 5-year follow up.

1. Introduction

Epithelioid hemangioendothelioma (EHE) is an uncommon vascular tumor that grows slowly and has a tendency to local recurrence. It develops in the limbs and less frequently in the head and neck region. Since its first description by Weiss and Enzinger in 1982 [1], very few cases have been reported in the orbit and/or the eyelid. To the authors' knowledge, only 7 cases of orbital EHE have been reported so far: 4 in the bone [2–5] and 3 in the soft tissues [6–8].

The authors of this article report a new case of EHE occurring in the right upper eyelid.

2. Case report

A 55-year-old male patient with a personal medical history of diabetes was referred to the first author of this paper in 2009 with a chief complaint of a growth in his right upper eyelid. The patient's surgical history showed that he underwent a surgical excision of a similar tumor in 2003. The tumor had been growing slowly in his right upper eyelid for two years before he decided to consult a doctor. There was no functional expression of its development. The histological examination of the specimen found then a benign fibrohystiocytic mesenchymal tumor that was incompletely removed. In 2006, the patient underwent a similar surgery and the histological findings were similar.

Three years later, the appearance of similar symptoms along with a right ptosis (Fig. 1) urged the patient to present in the Department of Plastic Surgery. The tumor was found to be extended to the internal half of the right upper eyelid. The palpation found a homogenously firm and almost immobile subcutaneous tumor. It extended to the upper palpebral conjunctival fornix. Ocular motility and palpebral sensitivity did not seem affected. The eye examination found nothing peculiar, except for a bilateral chorioretinal atrophy. The regional and general physical examination found neither regional lymph nodes nor similar lesions in the limbs.

Orbital CT scan and MRI revealed a multilobal tumor (Fig. 2) extended in the orbital space, outside the orbital cone. The globe, the right medial rectus muscle and the bone were exempt of any lesion (Figs. 3 and 4).

An incisional biopsy was then performed. It unveiled the bleeding potential of the tumor. The histological findings found a proliferation made of multicell layers. The cells showed a poorly limited cytoplasm with round or elongated nuclei and rare cytonuclear atypias. Intracytoplasmic vacuoles wrapping one or more red blood cells were found inside some cells. No mitotic figures were to be found. Immunohistochemical staining was positive for CD34 (Fig. 5) but negative for CD31. The pathologist's conclusion matched with the diagnosis of EHE.

Eventually, the patient was admitted to the operation theatre where he underwent a surgical excision of the tumor through an anterior orbitotomy. The specimen was firm, hemorrhagic and

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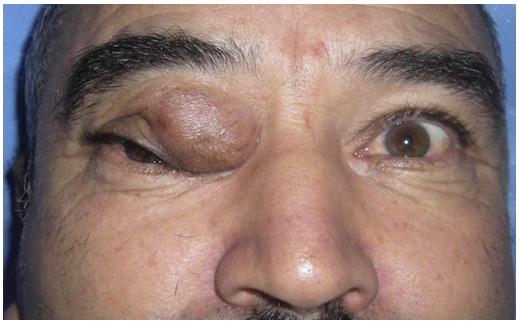

Fig. 1. Superomedial swelling of the right upper eyelid, associated to ptosis.

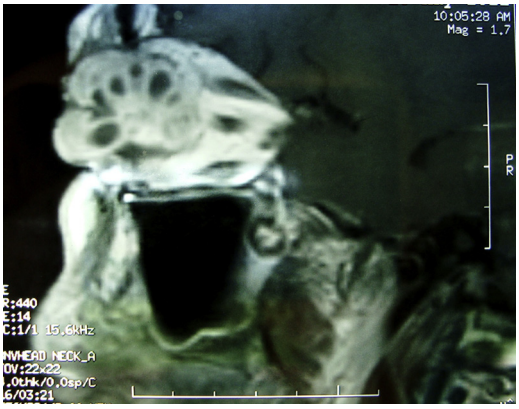

Fig. 2. RMI: sagittal T1-weighted image showing a multilobal mass of the orbit.

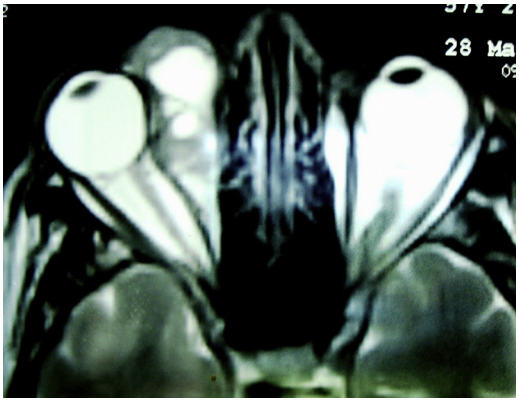

Fig. 3. RMI: axial T2-weighted image showing intraorbital high signal mass. No evident signs of muscle or ocular invasion.

sticking tightly to the skin and the conjunctiva. The histological study of the specimen confirmed the diagnosis of EHE. The excision margins were all within normal tissue. No recurrence was detected 5 years after the surgery.

3. Discussion

Epithelioid hemangioendothelioma (EHE) is an uncommon vascular tumor that was described for the first time by Weiss and Enzinger [1] in 1982. Its malignancy potential is intermediate and essentially related to its location. EHE has been reported in lungs, liver, spleen, bone, lymph nodes, brain and meninges [2,9]. When EHE occurs in soft tissues, it usually involves the limbs and appears as an indolent, solitary or multifocal tumor. Its occurrence in the head and neck region is very rare.

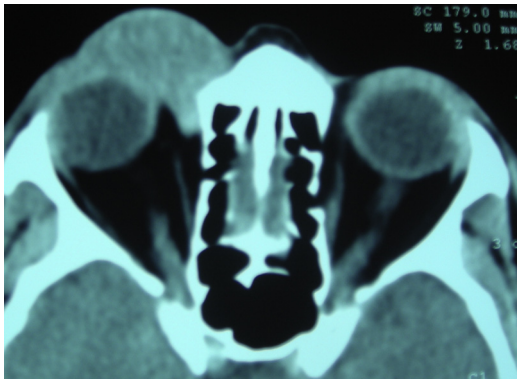

Fig. 4. CT scan: axial cut showing an intraorbital heterogenous process involving medial orbital soft tissues with no signs of bone lysis.

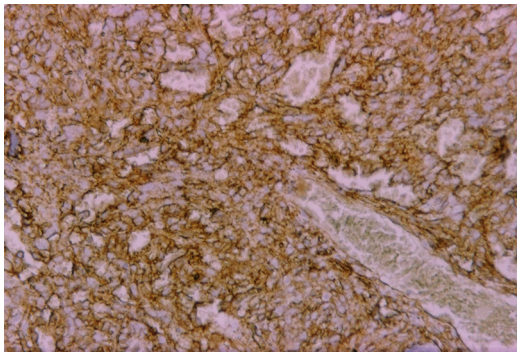

Fig. 5. Immunohistologic staining: tumor cells are positive for CD34.

Cervicofacial EHEs are divided into two entities:

- bony EHE;
- EHE of the soft tissues.

Symptoms depend mainly on the site and the size of the tumor. Because of its episodic surgings, the tumor produces a masse effect and a compression on its neighboring organs, without tissular invasion [3].

The most common vascular tumors of the orbit include:

- capillary hemangioma;
- cavernous hemangioma;lymphangioma;
- hemangiopericytoma [10].

To the authors' knowledge, only 7 cases of orbital EHE have been reported so far. In 4 cases, the tumor was located in the bone [2–5]. The 3 other cases involved the orbito-palpebral soft tissues: the lachrymal gland in one case [6], the superior tarsus in the second case [7] and the internal superior part of the orbital content in the third case [8].

Clinical features of the EHE are not specific to the disease. Symptoms like: pain, exophtalmia and ptosis depend on the tumor site and the level of compression it exerts on the neighboring tissues. CT scan and MRI are the main imaging techniques used for orbital tumors. They reveal the vascular nature of the tumor and assess the masse effect it exerts on the neighboring structures. The study of the 7 cases reported in the literature shows that the bony EHE are characterized by an obvious destruction of the bone. In the soft tissues EHE however, the tumor grows and produces a masse effect on the neighboring tissues without ever invading them.

Pathology is the key to diagnosis. The tumor is made of short strands or cords or small clusters of epithelioid round or outspread

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