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Case Report

Orbital solitary fibrous tumor: A painless mass after a dacryochystorhinostomy

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Abstract

The solitary fibrous tumor (STF) is a rare spindle cell tumor that most often occurs in the pleura, mediastinum and other serosal sites, but it can be found anywhere. The authors present a case report of a 35-year-old male who was submitted to an eventless left external dacryocystorhinostomy (DCR) due to epiphora. During the surgery no tumors or anomalous tissues were visualized. Five months later the patient presented a painless mass at the lower medial quadrant of the left orbit, near the lacrimal fossa. A computed tomography, a magnetic resonance imaging and an orbital ultrasound were performed. A round, very vascularized and highly contrast enhanced mass was found. An anterior orbitotomy was done. Histological aspects and immunohistochemical markers were consistent with STF. The tumor was not visualized during the DCR. The authors think that the growth of the tumor and the weakening of the tissues in the medial canthus after the DCR allowed the tumor to move anteriorly within the orbit.

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Introduction

The solitary fibrous tumor (SFT) is a rare spindle cell tumor that most often occurs in the pleura, mediastinum and other serosal sites.[1–5] The SFT, the hemangiopericytoma, the giant cell angiofibroma, and the giant cell fibroblastoma are considered to be within the same disease spectrum, due to overlapping of morphologic and immunophenotypic findings.[2] In the orbit it is rare and almost always extraconic, but it can be found anywhere.[6] It is a slow growing tumor with a mostly benign behavior. The gold standard treatment is surgery, however Nafiseh Hashemi et al.[7] treated a SFT with embolization followed by surgery.

Case report

A healthy 35-year-old man was observed in our department due to epiphora of the left eye. The lacrimal probing was done and confirmed nasolacrimal duct obstruction. The patient was submitted to an eventless left external dacryocystorhinostomy (DCR). During the surgery no tumors or anomalous tissues were visualized.

Five months later, the patient presented a painless mass at the lower medial quadrant of the left orbit, near the lacrimal fossa, without inflammatory signs or epiphora. The visual acuity was 20/20, indirect ophthalmoscopy and ocular motility were normal. An orbital computed tomography revealed a round lesion with intense and homogeneous contrast

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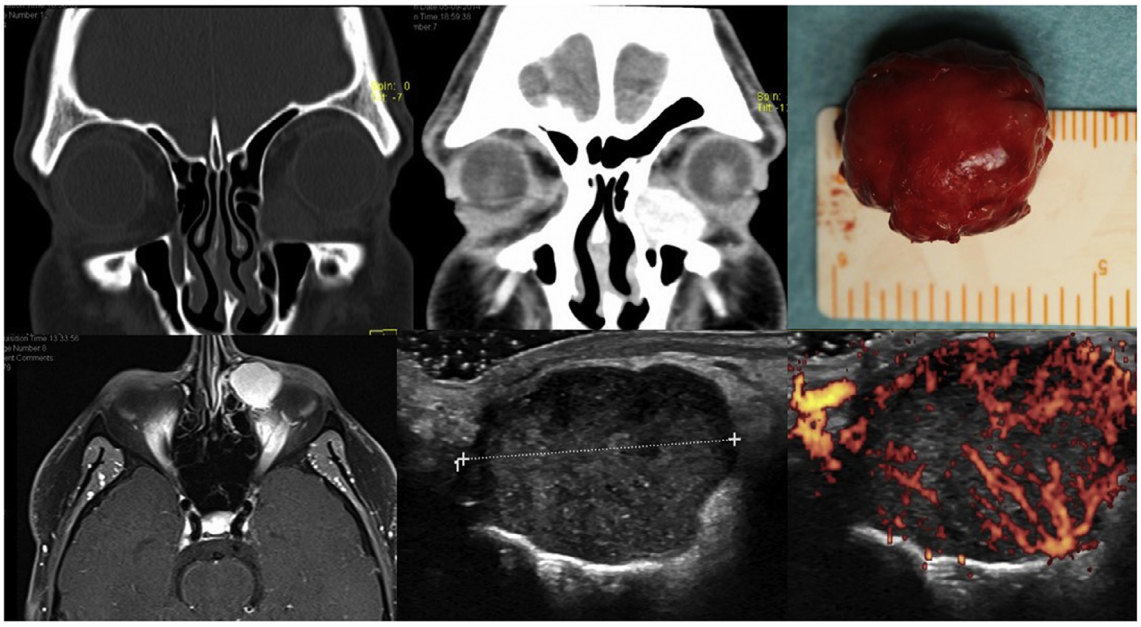

Fig. 1. Top- CT and STF; Botton: MRI and US.

enhancement, with mass effect at the inferomedial wall of the orbit (Fig. 1). The diagnosis of cavernous angioma or neoplasia were considered. The orbital magnetic resonance imaging showed a round lesion with hyposignal on T1 and low signal intensity on long TR, with intense and homogeneous contrast enhancement (Gd-DTPA). An inflammatory lesion or a non-aggressive neoplasia were considered as differential diagnosis. An orbital ultrasound revealed a solid nodular lesion, homogeneous, hypoechoic and highly vascularized without the usual features of a hemangioma or other vascular malformation and compatible with a giant cell angiofibroma.

An anterior orbitotomy with transconjunctival approach was performed and a round, well encapsulated, elastic, red lesion was removed.

The lesion weighted 4 g, and had the following dimensions: 2,5 × 1,6 × 1,4 cm. Histologically, it was a highly vascularized lesion with cancer cells sometimes ovoid, sometimes fusiform, inside a collagenous stroma with slight pleomorphism and marked capillary vascularization in a pattern compatible with a mesenchymal neoplasia. The immunohistochemistry (Fig. 2) was CD 34+, Bcl2+, CD117−, S100−, SMA−.

In the following three years there was no recurrence of the lesion.

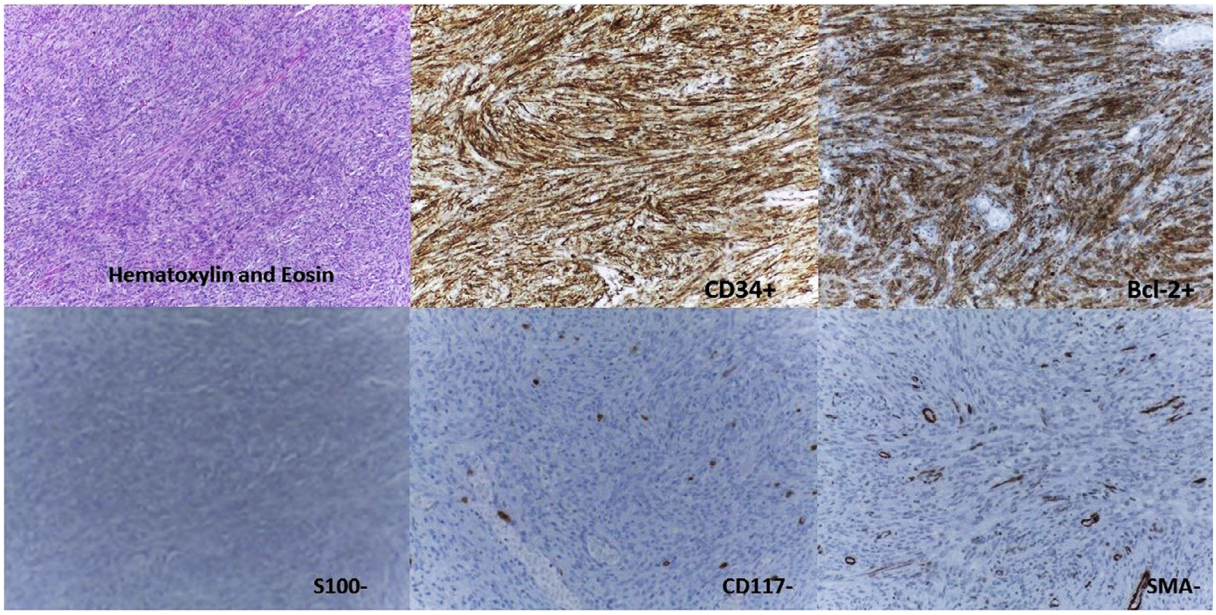

Fig. 2. Histology and immunohistochemistry.

Discussion and conclusion

Clinical examination, imaging, and histology are nonspecific in the evaluation of the SFT of the orbit.[4,8] Therefore, immunohistochemical markers are fundamental for the diagnosis.[6] The CD34 is considered the most reliable marker for orbital STF, but it is nonspecific.[9,11] The expression of Bcl-2 is very high in STF and negative in most malignant mesotheliomas.[9] Thus, the combination of these two immunohistochemical markers increase the diagnostic accuracy. In our patient, the histological pattern and the immunohistochemistry (CD34+, Bcl-2+) are consistent with a STF of the orbit.

In fact, the peripheral nerve sheath tumors are S100+,[6] the leiomyosarcoma and mesothelioma are CD34−, so both are excluded. The GISTs may be CD34 and SMA positive or negative, depending on the site of the tumor in the gastrointestinal tract,[10] but are CD117+ and Bcl-2+, so are also excluded.

An orbital STF is a slow growing, indolent and mostly nonaggressive tumor.[5] The proliferation of cells may result from the high expression of Bcl2, a potent inhibitor of apoptosis,[9], and not from cell mitosis which are scarse in SFT.

In this patient the tumor was not visualized during the DCR. Therefore, we think that the growth of the tumor and the weakening of the tissues in the medial canthus after the DCR allowed the tumor to move anteriorly within the orbit. The treatment of the orbital SFT consists in complete surgical removal[12] which was performed. No recurrences occurred within the three year follow-up.

Conflict of interest

The authors declared that there is no conflict of interest.

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