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Case Report

Orbital solitary fibrous tumor☆

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ABSTRACT

We present the case of a 46-year-old man who complained of progressive right lower eyelid swelling associated with diplopia when looking down. A nontender mass over the right lower palpebral conjunctiva with vessel engorgement and limited downward gaze were found. Magnetic resonance imaging (MRI) demonstrated a well-defined nodular lesion occupying the inferior area of the orbit. After surgery, a pathological examination revealed that the lesion was a well-encapsulated fibrous tumor composed of spindle cells with a whorl-like arrangement and collagen formation. Immunohistochemical staining was positive for vimentin, CD34, and CD99. The Ki-67 level was relatively low, diagnostic of a solitary fibrous tumor. Our case showed a typical presentation of an orbital solitary fibrous tumor with slow progression, MRI images of a homogenous isodense lesion in T1, heterogeneous isodense lesion in T2, and positive vimentin, CD99, and CD34 on cytological analysis. This case shows that a combination of history, radiology, and pathology results is crucial to differentiate soft-tissue tumors in the orbit.

1. Introduction

A solitary fibrous tumor is an uncommon spindle cell neoplasm of mesenchymal origin, arising from the pleura, pericardium, and mediastinum. Extraserosal locations such as the lungs, liver, and nasal sinus have also been reported. Two cases of orbital solitary fibrous tumors (OSFTs) was first described in 1994 [1]. The clinical manifestations, radiological findings, and pathological features of OSFT in different ethnic groups have also been discussed in the literature [2]. We herein report a case of an OSFT with a typical presentation in a Chinese patient.

2. Case report

A 46-year-old man complained of progressive right lower eyelid swelling for 1 year accompanied by diplopia on downward gaze. On examination, a nontender mass over the right lower palpebral conjunctiva with vessel engorgement and limited extraocular muscle movement during downward gaze were noted. Magnetic resonance imaging (MRI) demonstrated a 2.1 cm × 1.9 cm × 2.2 cm well-defined nodular lesion beneath the right lower eyelid (Fig. 1).

The patient underwent excision of the orbital tumor. A well-encapsulated 2.5 cm × 2.0 cm × 1.8 cm mass with adhesion to the inferior oblique and inferior rectus muscles was found (Fig. 2), with no local invasion to the adjacent tissue. A microscopic analysis revealed it to be a well-encapsulated fibrous tumor composed of spindle cells with a whorl-like arrangement and collagen formation. Immunohistochemical staining was positive for vimentin, CD34, and CD99, and focally positive for Bcl-2 and S100. The level of Ki-67 was relatively low (Fig. 3), suggesting a solitary fibrous tumor. After the operation, the eyelid swelling and diplopia resolved.

3. Discussion

The OSFT condition has been reported in patients at different ages, but mostly in the fourth decade of life, as in our patient [2,3]. It is relatively rare in children [4,5], and is not sex-specific [2]. It appears to occur less frequently in those of Chinese ethnicity, and only four cases have been reported in Taiwan so far [6–8]. Common clinical presentations are eyelid swelling, diplopia, and misalignment, according to the location of the tumor [1]. Tumors in the lacrimal apparatus such as the lacrimal gland and sac leading to proptosis or epiphora have been reported [9–11].

Radiological imaging is important in preoperative examinations. Imaging findings of OSFT on computed tomography (CT) and MRI provide some clues. The lesions tend to be oval, well-defined, and isodense in the extraocular muscles on CT. Marked enhancement

Conflict of interest: none.

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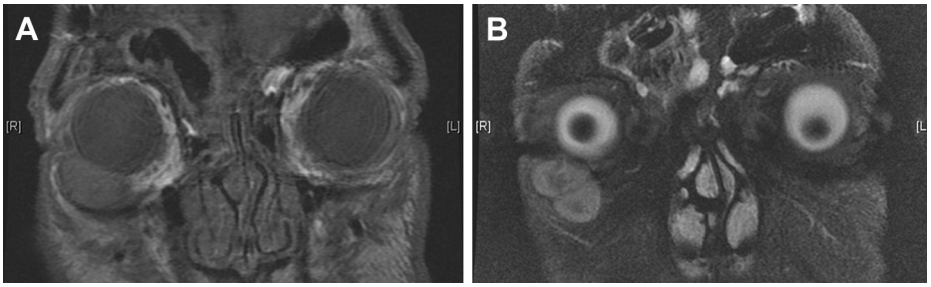

Fig. 1. (A) A well-encapsulated tumor displays homogeneous isodensity in an MRI T1 sequence without gadolinium. (B) A coronal noncontrast MRI T2 sequence demonstrates a heterogeneous isodense tumor. MRI = magnetic resonance imaging.

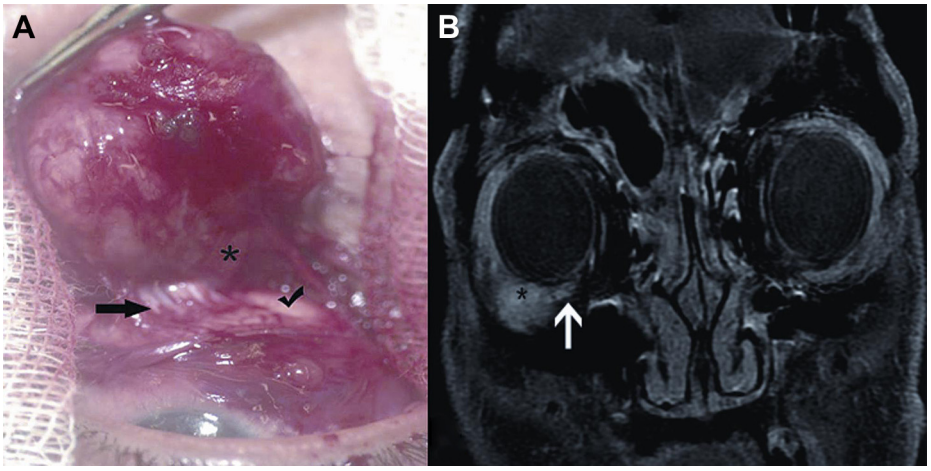

Fig. 2. (A) Intraoperatively, a well-encapsulated mass without adhesion to adjacent tissue is seen. The mass with adhesion to the inferior rectus muscle (arrow), inferior oblique muscle (asterisk), and sclera (check mark) is shown. (B) Tumor adhesion to the inferior rectus muscle on magnetic resonance imaging; inferior rectus muscle (arrow) and tumor (asterisk).

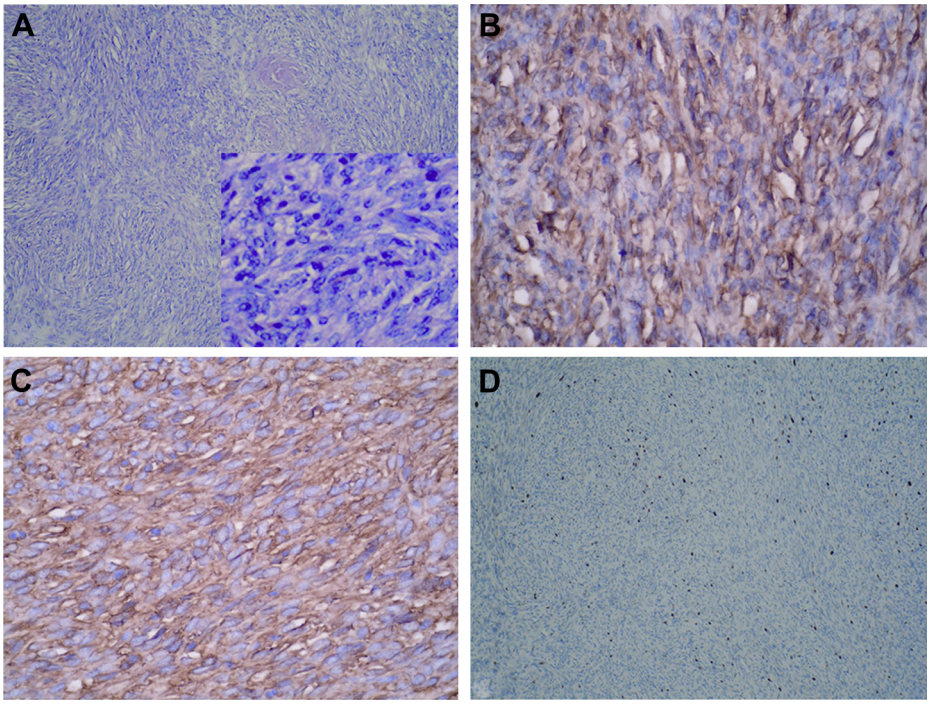

Fig. 3. (A) Hematoxylin and eosin staining shows proliferation of spindle cells with collagen in the center (original magnification: 100×; inset magnification: 400×). (B) The tumor cells are positive for CD99 (++) and (C) CD34 (+++) (immunohistochemical stain; original magnification: 400×). (D) The Ki-67 level is relatively low (3%) (immunohistochemical stain; original magnification: 40×).

has been noted in contrast series [12]. The value of CT is in detecting bone involvement rather than involvement of the soft tissues of the orbit [13], for which MRI is more useful. On T1-weighted images, tumors are homogenous isodense to gray matter, whereas they are heterogeneous isodense or hypodense on T2-weighted images [14,15]. Although a few cases have shown heterogeneous hyperdensity or a cystic appearance, this is a key to differentiating OSFT from schwannomas [16,17]. Metastatic disease, the most common orbital tumor in adults, usually shows nonspecific features on CT and MRI, except for multisite involvement [18].

Pathological findings play the most significant role in distinguishing OSFT from other neoplasms such as benign fibrous histiocytoma, schwannomas, hemangiopericytomas, and giant cell angiofibromas/fibroblastomas. Microscopically, solitary fibrous tumors are composed of round to spindle cells with focal staghorn vasculature [19]. The key to diagnosis lies in immunohistochemical reactions, as these lesions are positive for vimentin, CD34, Bcl-2, and CD99, but negative for desmin, epithelial markers (cytokeratin), vascular markers (factor VII-related antigen), neural markers (S100 protein), and muscle markers such as muscle-specific actin and smooth muscle actin [20,21]. However, focal positivity for S100 was noted in our case and in other reports. Malignant OSFT is uncommon. The histological criteria for malignancy are hypercellularity, moderate-to-marked cytological atypia, necrosis, and more than four mitoses per 10 high-power field or an infiltrative margin [6,20]. Malignant OSFT may show a loss of CD34 immunoreactivity [22]. In our case, there was no evidence of malignancy as there was no atypia or necrosis, only a few mitosis and a low Ki-67 level (<3%). Although local invasion and recurrence are possible, the prognosis is generally good [23–25]. Chemotherapy and radiotherapy after excision have been used successfully [26,27]. Parrozzani et al [28] reported a case of a solitary fibrous tumor of the orbit with intracranial invasion in which local recurrence and systemic metastasis occurred 3 years after surgery. A biphasic tumor pattern of spindle cells and an epithelioid component were noted. This may therefore be a risk factor for a poor prognosis.

In conclusion, OSFT should be evaluated in combination with clinical manifestations, imaging findings, and histological features. Our case was diagnosed because of sufficient immunohistochemical evidence. After complete excision, the recurrence rate is low and further local treatment is unnecessary. More cases are needed to investigate the characteristics in a specific population.

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