Recurrent Orbital Solitary Fibrous Tumor in a 12-Year-Old

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Keywords

Solitary fibrous tumor · Recurrent pediatric orbital tumor · Excision

Abstract

Purpose: To report the clinical features and pathology of orbital solitary fibrous tumors and present a case of a recurrence in a 12-year-old boy. **Methods:** Case Report. **Results:** A 12-year-old boy presented to our institution with double vision in upgaze for 6 months. Examination revealed 3 mm of left-sided proptosis, hypoglobus, and –3 impairment of supraduction. Magnetic resonance imaging of the orbits showed an avidly enhancing mass within the superolateral aspect of the orbit. Biopsy revealed a solitary fibrous tumor with STAT6 and CD34 positivity. Positron emission tomography-computed tomography was negative for metastatic disease. The tumor underwent excision. However, 4 months postoperatively, imaging revealed changes consistent with recurrence. A repeat lateral orbitotomy with orbital rim marginotomy was performed with complete gross excision of the tumor. The patient remains tumor-free 22 months after reoperation. **Conclusion:** Orbital solitary fibrous tumor rarely occurs in the pediatric population. Despite initial com-

plete gross tumor excision, this case represents the youngest individual with a recurrence to the authors' knowledge. This case of rapid recurrence may have been due to tumor seeding at the time of initial biopsy or other factors. This case increases the known spectrum of orbital solitary fibrous tumors.

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Solitary fibrous tumors (SFTs), first described in 1931 by Klemperer and Rabin [1], represent rare, generally benign, mesenchymal neoplasms that most commonly involve the pleura. They occasionally show signs of malignant behavior, such as adjacent tissue invasion, recurrence, or metastasis [2]. Bernadini et al.'s review of the literature [3] found a recurrence rate of 19% for orbital SFTs.

SFTs in the pediatric patient population are very rare as the typical mean age of presentation is 41.6 years [4]. A review of the literature in 2003 described a total of 48 patients with orbital SFTs, only 3 of which were pediatric cases [5]. The youngest reported case occurred in a 5-year-old and two other pediatric recurrences have been reported in a 14-year-old girl and 15-year-old girl, respectively

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Fig. 1. Pediatric patient with left hypoglobus, proptosis, and elevation deficit.

Fig. 2. Coronal MRI (T1-weighted) showed an avidly enhancing mass within the superolateral aspect of the left orbit.

[6–8]. Patients with these tumors typically present with unilateral painless proptosis, eyelid swelling, a palpable mass, tearing, and/or blepharoptosis [3]. On magnetic resonance imaging (MRI), SFTs appear homogeneously isointense to gray matter on T1-weighted images, whereas they give an iso- or hypointense signal on T2-weighted images, have marked enhancement, and have a time-intensity curve with a washout pattern [9]. Histological characteristics include thick and often keloid-like bands of collagen, alternating areas of hyper- and hypocellularity, and a hemangiopericytoma-like pattern of vascularity. Malignant histologic features include hypercellularity, demonstrating at least focally moderate to marked cytological atypia, tumor necrosis, increased mitotic rate (greater than 4 per 10 high-power fields), and infiltrative margins [10]. However, imaging and surgical findings may more accurately predict aggressive behavior [11]. Although these tumors generally follow an indolent and benign course, a 2013 review of orbital SFTs found that 20 of 84 demonstrated recurrence; however, one tumor did exhibit metastasis. Initial complete resection, rather than histologic features, represents the most important prognostic factor [12].

Case Report

In December 2013, a 12-year-old boy presented to our institution with a 6-month history of progressively worsening swelling of the left eye associated with headaches. He denied any significant

past medical history, periocular trauma, or history of amblyopia. On external examination, the left eye was observed to have hypoglobus and proptosis. Best corrected visual acuity was 20/20 in the right eye and 20/70 in the left eye. Hertel measurements were 15 mm OD and 18 mm OS with a base of 96 mm. The patient had difficulty with upgaze and a –3 impairment of supraduction (Fig. 1). MRI of the orbits revealed a lobulated, avidly enhancing mass within the superolateral aspect of the left orbit, measuring $2.7 \times 2.6 \times 2.3$ cm. The mass was predominantly extraconal, but also extended intraconally and exhibited a mass effect on the inferomedially deviated left optic nerve and globe (Fig. 2). Biopsy revealed a spindle cell neoplasm with myxoid to collagenous background and extensive vasculature with varying degrees of perivascular hyalinization. The tumor cells were monomorphic with oval nuclei, darkly staining chromatin, inconspicuous nucleoli, and scanty eosinophilic cytoplasm. Mitotic figures, necrosis, and histologic features of malignancy were absent. Immunohistochemistry revealed tumor cells positive for CD34 and STAT6 and negative for desmin, cytokeratin AE1/AE3, p63, epithelial membrane antigen, and S-100 protein, consistent with the diagnosis of SFT (Fig. 3). Systemic workup, including a bone scan and CT of the chest, abdomen, and pelvis was unremarkable. The patient underwent a left lateral orbitotomy with complete gross excision of the lesion. Postoperative imaging was not done at this time. Surgical pathology revealed morphology similar to the previous biopsy.

Four months later, the patient presented with recurrent proptosis. Imaging showed a heterogeneously enhancing post-septal mass along the superolateral left orbit, measuring $2 \times 1.9 \times 2.8$ cm with necrotic changes, consistent with recurrence. A left lateral orbitotomy with marginotomy was utilized for repeat excisional biopsy (Fig. 4). The specimen showed a predominant myxoid pattern, STAT6 immunoreactivity, and more mitotic activity than seen previously, now up to one mitotic figure per 10 high-power fields. No necrosis, increased cytologic atypia, or overt sarcomatous progression was identified.

Fig. 3. **a** Spindle cell neoplasm with myxoid background in a “patternless pattern” with extensive vasculature (hematoxylin-eosin, $\times 200$). **b** Predominant nuclear STAT6 immunostaining positivity ($\times 200$).

Fig. 4. Postoperative picture of the patient following lateral marginotomy and complete excision of the tumor.

Three-month follow-up revealed improvement in visual acuity of the left eye correctable to 20/20, mild left enophthalmos, and no impairment of supraduction. Though no repeat surveillance for metastatic disease was performed, CT and MRI imaging of the orbits continued to show no signs of disease. The patient remained asymptomatic with no sign of recurrence 22 months after the second surgery.

Discussion

SFTs are generally benign spindle cell neoplasms of mesenchymal cell origin that can arise not only in the pleura, but also in extrapleural sites such as the orbits [5]. Other orbital tumors for differential consideration include histiocytomas, giant cell angiofibromas, and hemangiopericytomas. Debate exists about whether these tumors represent variants of orbital SFTs [13]. The overlap of clinical and histopathologic characteristics between these tumors requires additional workup with immunohistochemistry. In our case, not only were the clinical and histopathologic features consistent with the WHO classification for orbital SFTs, but the tumor also expressed CD34 positivity, which has classically been used to differentiate SFTs from other tumors [10]. Furthermore, a 2015 study demonstrated higher sensitivity and specificity with nuclear STAT6 and cytoplasmic ALDH1 expression [14]. The tumor in this case exhibited STAT6 positivity as well. Complete resection of the tumor, rather than histological characteristics, represents the most important

prognostic factor and necessitates close follow-up [12]. If the initial excision is incomplete, the recurrent tumor tends to spread into surrounding tissues and bone, rendering a second excision much more difficult [3].

This case represents the youngest reported case of an orbital SFT that recurred following surgical resection. Although uncommon, orbital SFTs should be included in the differential diagnosis of orbital tumors in the pediatric population. Despite complete gross resection, these tumors may recur and require even more aggressive wide excision.

Statement of Ethics

This investigation was compliant with the Health Insurance Portability and Accountability Act and adhered to the tenets of the Declaration of Helsinki. Informed consent was obtained for this case report. Institutional Review Board approval was not applicable.

Disclosure Statement

The authors have no financial disclosures to report. The authors have no financial or proprietary interests related to this case report.

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