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Solitary fibrous tumor of the orbit with growth during pregnancy: a case report

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Abstract

We report the case of an anterior orbital tumor in a young woman that enlarged during pregnancy. The mass was excised and found to be a spindle cell tumor with immunohistochemical reactivity consistent with a solitary fibrous tumor, a rare entity in the spectrum of fibroblastic mesenchymal tumors. The tumor was strongly positive for the progesterone receptor, consistent with its clinical growth during the antenatal and postnatal periods. To our knowledge, a primary orbital tumor with these characteristics has rarely been reported in the literature.

Introduction

Fibroblastic tumors of the orbit are rare neoplasms with approximately 80 cases reported to date in the literature and conflicting evidence regarding their pathologic diagnosis.¹ We report the case of a patient referred to the oculoplastics service for evaluation of an orbital mass which was found on pathologic analysis to represent a mass of spindle cells differentiated by immunohistochemical analysis as a solitary fibrous tumor. Collection and evaluation of protected patient health information was HIPAA-compliant.

Case Report

A 32 year old woman with a history of a recent uncomplicated pregnancy presented with a 1 year history of a progressively enlarging painless right upper eyelid lesion associated with occasional tearing and mucoid discharge. She denied diplopia, photopsias, decreased vision or headaches. She first noticed the lesion during the second trimester of her pregnancy and reported that it had continued to grow following an uncomplicated delivery at term. There was no personal history of malignancy, ocular surgery, trauma, ear or sinus infections and she was not using any ocular medications. Family history was notable for 2 aunts with *BRCA1/2*-negative breast cancer. She reported a 12 year smoking history, did not drink alcohol, denied illicit drug use, and reported no drug allergies.

On examination, best corrected visual acuity was 20/20 in each eye. In the right anterior orbit, there was a 1 cm by 1 cm circumscribed palpable mass in the superior medial aspect with concomitant mechanical blepharoptosis and deformation of the eyelid crease. There

was no overlying erythema, discharge, tenderness, or associated lymphadenopathy. Levator function was intact and symmetric for both eyes and there was no lagophthalmos or proptosis. The remainder of the ocular examination was within normal limits.

Magnetic resonance imaging of the orbit with and without gadolinium was obtained and revealed an extraconal $2.2 \times 1.4 \times 2.2$ cm enhancing mass with internal cystic components within the superomedial aspect of the right orbit (Figure 1). The mass abutted the right superior oblique muscle with lateral displacement of the right medial rectus and globe.

An anterior orbitotomy was performed with removal of the mass. Light microscopy of the excised mass showed a circumscribed tumor comprised of ovoid to plump spindle-shaped cells with moderate eosinophilic cytoplasm and ovoid to elongated vesicular nuclei with inconspicuous nucleoli (Figure 2A). Up to 3 mitoses were seen per high power field. Background stroma was fibrocollagenous and richly vascular with the presence of irregular and dilated vessels, some of which had a staghorn appearance. Nuclear atypia was minimal. The tumor cells exhibited patchy positivity for CD34 (Figure 2B) and were negative for CD31, SMA, and S100. The tumor cells exhibited diffuse cytoplasmic reactivity for Bcl2 and diffuse nuclear immunoreactivity for STAT6 (Figure 2C). A strong expression of progesterone receptor (PR) was seen in these tumor cells (Figure 2D). However, fewer than 5% of cells expressed estrogen receptor (ER). Ki67 labelling index was less than 5%. SS18/SSX1 and SS18/SSX2 fusion transcripts were not detected by RT-PCR.

The patient had an unremarkable postoperative course and was referred for an oncological workup. No further intervention was required.

Discussion

We report here a case of a progesterone receptor-positive tumor consistent with a solitary fibrous tumor. To our knowledge, a primary orbital tumor with the morphological and immunohistochemical profile described herein has seldom been reported in the literature.

The differential diagnosis of circumscribed orbital neoplasms with fibrous and vascular differentiation is broad but includes solitary fibrous tumor, hemangiopericytoma, giant cell angiofibroma, and fibrous histiocytoma. The defining feature of solitary fibrous tumor, irrespective of tumor site and histologic grade, is strong and diffuse immunoreactivity against the human hematopoietic progenitor cell antigen, CD34,^{2,3} a feature not present in the tumor cells in this report. Occasionally, hemangiopericytomas may feature CD34 immunoreactivity with weak intensity and patchy distribution, but the absence of the characteristic staghorn sinusoidal blood vessels on histopathology in this case excludes that diagnosis. The pattern of CD34 positivity observed in this tumor resembled that which can be occasionally observed in smooth muscle tumors such as leiomyomas and leiomyosarcomas, but this lesion was not immunoreactive to SMA. Neither multinucleated giant cells nor a focal storiform architecture were observed, excluding giant cell angiofibroma and fibrous histiocytoma. It has been reported that STAT6 expression is a useful diagnostic marker of solitary fibrous tumors and reliably can distinguish these

variable neoplasms from mimics.⁴ Indeed, in this case, the strong expression of STAT6 differentiates the neoplasm as a solitary fibrous tumor.

A surprising characteristic of this tumor was its clinical growth during pregnancy, consistent with its immunoreactivity to the progesterone receptor. Elsewhere a progesterone receptor-positive orbital tumor that exhibited accelerated growth during pregnancy had been reported.⁵ In that report the tumor was consistent with a schwannoma; in this case, however, S100 staining was negative. The case of an orbital intramuscular hemangioma enlarging during pregnancy has also been described,⁶ as well as that of a solitary fibrous tumor of the orbit,⁷ but the immunohistochemical and histological profile of the lesion described here is distinct from that described in those reports.

Debate in the literature exists regarding the classification of hemangiopericytoma, solitary fibrous tumor, giant cell angiofibroma, and fibrous histiocytoma, with some consideration that these all represent tumors with a common lineage and should be classified as variants of solitary fibrous tumors.^{1,8,9} There is nevertheless considerable diversity in their behavior, and further exploration is needed to characterize well the possible malignant potential¹⁰ and growth behavior of these rare tumors. Indeed, an earlier study suggested that some CD34+ tumors may exhibit progesterone receptor positivity.¹¹ The case described herein highlights not only the overlapping histological and immunological profiles of these rare fibrovascular tumors, but enhances our understanding of the biological behavior of solitary fibrous tumors.

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Fig 1.
Fibrous tumor right orbit

Fig 2.
Fibrous circumscribed tumor