Fairooz P. Manjandavida, Bangalore, India
Carol L. Shields, Philadelphia, USA

The second time around

Case

A 42-year-old Caucasian woman with 2 mm of left eye proptosis for 3 months was referred for a recurrent intraconal mass. She was otherwise healthy with ocular history of LASIK in both eyes (OU). On examination, best-corrected visual acuity was 20/25 OU. Intraocular pressure, extraocular movements, optic nerve function, and dilated fundus examination were normal. Three years prior she had undergone excision of an orbital mass [Fig. 1a], demonstrating spindle cell morphology, consistent with solitary fibrous tumor (SFT). Repeat magnetic resonance imaging of the orbit showed interval recurrence of an irregular hyperintense soft tissue mass on T1-weighted images, extending to the orbital apex and optic nerve [Fig. 1b].

What Is Your Next Step?

A. Corticosteroid injection
B. Chemotherapy
C. Surgical re-excision
D. Observation

Findings

The primary excision showed a 22 × 18 × 15 mm encapsulated mass [Fig. 1c], composed of spindle cells with mildly pleomorphic and focally overlapping nuclei [Fig. 1d], with five mitotic figures in 10 high-power fields (HPFs). Repeat excision showed frankly malignant features including atypical mitoses [Fig. 1e, yellow arrow], marked nuclear pleomorphism, focal absence of STAT 6 expression on immunohistochemistry [Fig. 1f, absence of expression black arrow, retained expression white arrow], and positive margins. Residual tumor at the left orbital apex could not be further excised without visual compromise, so external beam radiotherapy of 70 Gy was administered. At 6 years of follow-up, best-corrected visual acuity was counting fingers secondary to cataract and radiation maculopathy. There was no further tumor recurrence.

Diagnosis

Malignant Solitary Fibrous Tumor with Foci of De-Differentiation, Recurrence.

Correct Answer: C

Discussion

Solitary fibrous tumor is a rare mesenchymal tumor, commonly affecting pleura and less often the orbit.[1] In the orbit, this mass presents with unilateral, slowly progressive proptosis in middle-aged adults, appearing circumscribed on orbital imaging.[1] Strong and diffuse STAT 6 immunoreactivity is highly sensitive for well-differentiated SFT, whereas loss of expression is observed in de-differentiated SFT with malignant potential.[2] Complete excisional biopsy and lifelong observation are advised. Incomplete excision can be associated with recurrence. Histopathologic features of malignancy in SFT include nuclear atypia, greater than four mitoses per 10 HPFs, necrosis, and infiltrative margins.[1-3] Adjuvant chemotherapy and radiotherapy have not yet been evaluated for efficacy.[3]

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Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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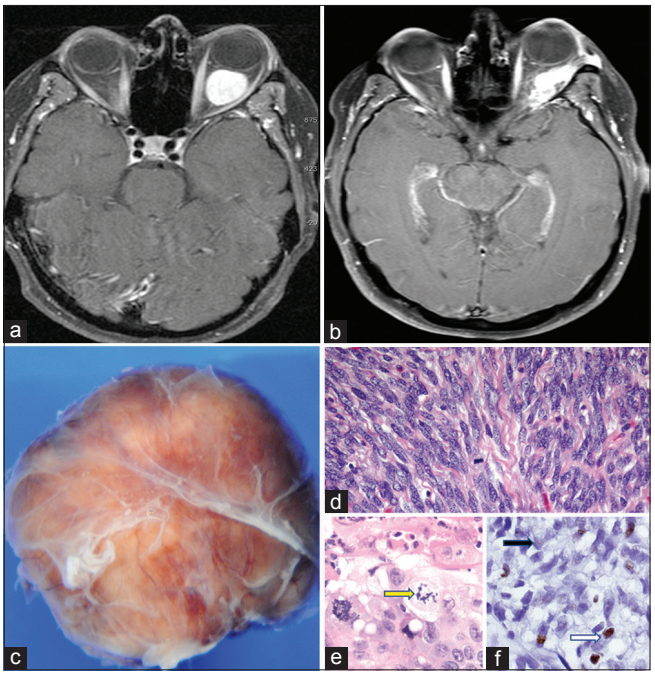

Figure 1: Three years prior, a left orbital mass had been excised with pathology consistent with solitary fibrous tumor (a). Repeat magnetic resonance imaging of the left orbit showed interval recurrence of an irregular hyperintense soft tissue mass on T1-weighted images extending to the orbital apex and optic nerve (b). The primary excision showed a 22x18x15 mm encapsulated mass (c) composed of spindle cells with mildly pleiomorphic and focally overlapping nuclei (d). Repeat excision showed frankly malignant features including atypical mitoses (e, yellow arrow), nuclear pleiomorphism and focal absence of STAT6 expression on immunohistochemistry (f absence of expression black arrow, retained expression white arrow)

Conflicts of interest

There are no conflicts of interest.

Li-Anne L Lim, Md K Hasanuzzaman, Carol L Shields

Department of Ocular Oncology, Wills Eye Hospital, Philadelphia, PA, USA

Correspondence to: Dr. Carol L Shields, Wills Eye Hospital, 840 Walnut Street, Philadelphia, PA 19107, USA.
E-mail: lianne@shields.md

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