Solitary Fibrous Tumor of the Sclera

Solitary fibrous tumor (SFT) is a spindle cell neoplasm of mesenchymal origin that typically arises from the pleura. Several extrapleural sites have also been described, including the orbit, lacrimal gland fossa, and lacrimal sac. We present the clinical, imaging, and histopathologic findings of an SFT arising from the sclera that manifested as an intraocular mass with extrascleral extension.

Report of a Case. A 50-year-old man reported a 1-month history of painless progressive vision loss in the left eye. Pertinent medical history was remarkable for smoking, hypertension, and hyperlipidemia. Best-corrected visual acuity was 20/20 OD and 20/400 OS with a relative afferent pupillary defect on the left side. There was no proptosis, and ocular motility was full. Anterior segment examination in the left eye was notable for a 13-mm subconjunctival mass in the inferotemporal bulbar region with overlying prominent conjunctival vessels. The other findings of the anterior segment examination, including gonioscopy and intraocular pressure measurement, were normal. Dilated fundus examination revealed a round amelanotic subretinal mass located at the 5-o’clock meridian posterior to the equator associated with overlying retinal detachment extending into the macula (Figure 1). A- and B-scan echography demonstrated a dome-shaped subretinal mass with low internal reflectivity and subtle vascularity associated with a scleral defect posterior to the tumor (Figure 2). Basal dimensions were 13.4 × 7.1 mm, and the apical height was 5.4 mm. Orbital magnetic resonance imaging demonstrated a well-circumscribed round mass in the inferotemporal quadrant of the left globe measuring 14 mm in maximal diameter between the intraocular and extrascleral components (Figure 3). The mass had its broadest dimension at the level of the sclera. The mass was predominantly isointense to gray matter on T1-weighted signal intensity and hypointense on T2-weighted images and displayed contrast enhancement. A metastatic workup did not show distant metastases or evidence of a primary extraorbital malignant neoplasm.

Although the referring clinical diagnosis was choroidal melanoma...
with extrascleral extension, the lesion was atypical for its rounded rather than “collar button” contour and epicenter at the scleral level. Management options included fine-needle aspiration biopsy, plaque radiation therapy, and enucleation. Because of the extensive scleral involvement, the possibility of full-thickness eye wall resection was not considered. In view of the possible scleral origin, the tumor was removed in its entirety via an enucleation of the left eye.

Gross examination of the specimen showed a firm, tan, round mass replacing the sclera in the inferotemporal quadrant of the globe (Figure 4). Histopathologic evaluation revealed a spindle cell neoplasm with no nuclear atypia and absence of mitotic activity (Figure 5). Immunohistochemical studies showed positive reactivity to CD34 (Figure 6) and negative reactivity to HMB45, S100, CD31, AE1/3 (anion exchanger 1/band 3), CD99, and smooth-muscle actin. The findings were consistent with the diagnosis of SFT arising from the sclera. The patient was free of local tumor recurrence and metastatic disease at 30 months of follow-up.

Comment. Solitary fibrous tumor has several important radiologic and histologic characteristics that are essential for establishing its diagnosis. The uniform immunohistochemical staining for CD34 supports a diagnosis of SFT and helps to differentiate this lesion from fibrous histiocytomas and fibrosarcomas, which are CD34 negative, as well as hemangioendotheliomas, which show weak and patchy CD34 staining only around blood vessels. Schwannomas are not only CD34 positive but also S100 positive, which is not a feature of SFT. Giant cell angiofibroma and giant cell fibroblastic stroma share similar immunostaining patterns; however, multinucleated giant cells and pseudovascular spaces are usually not observed in SFT.

The imaging studies in this case also provided several characteristic features that support the diagnosis of SFT. The echographic features of the lesion agree with published findings of orbital SFT, which typically show a well-outlined lesion of low and regular internal reflectivity attributed to the histologic correlate of densely packed spindle cells. In addition, the magnetic resonance imaging characteristics observed in this case are consistent with the imaging findings in SFT of the orbit. The typical findings on magnetic resonance imaging include a well-delineated enhancing mass lesion with intermediate T1-weighted signal intensity and low T2-weighted signal intensity. The low signal intensity on T2-weighted imaging is thought to correlate with the lack of tissue water density from the collagenous stroma of SFT. This fea-
ture is in contrast to other spindle cell tumors such as fibrous histiocytoma and hemangiopericytoma, which are usually hyperintense on T2-weighted images. Although the magnetic resonance imaging characteristics of SFT are similar to those of melanoma, the rounded appearance observed in our patient with its epicenter at the level of the sclera challenged the diagnosis of melanoma with extrascleral extension and raised the suggestion of a tumor arising from the sclera.

The clinical behavior of orbital SFT is usually benign, although some of these tumors have exhibited aggressive features such as infiltration of the orbital roof and optic nerve, as well as extraorbital extension. The treatment for SFT is en bloc excision because incomplete removal results in a higher recurrence rate and carries a potential for malignant transformation. Careful long-term follow-up after complete excision seems to be the standard of care.

To our knowledge, this case represents the first published report of SFT arising from the sclera. Although rare, SFT should be included in the differential diagnosis of intraocular and epibulbar masses.

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Uveitis Associated With Atypical Cogan Syndrome in Children

Typical Cogan syndrome1 involves nonsyphilitic interstitial keratitis associated with audiovestibular involvement that progresses to complete deafness within 2 years. Atypical Cogan syndrome occurs when sensorineural hearing loss is associated with ocular inflammation distinct from interstitial keratitis, such as uveitis, scleritis, conjunctivitis, or retinal vasculitis. We describe 3 cases of atypical Cogan syndrome that were undiagnosed for a long time. Awareness of this syndrome will aid in the diagnosis and subsequent treatment of this unusual entity. Cogan syndrome is described as typical when it appears in young adults with flare-ups of interstitial keratitis and sudden onset of Ménière disease–like attacks, which progress to sensorineural deafness within 2 years. The keratitis is usually bilateral peripheral subepithelial keratitis, which can progress to nummular lesions. A deep stromal keratitis can occur and vascularization of the cornea usually occurs. Early diagnosis is most important, as early treatment of cochlear symptoms with corticosteroids may prevent or lessen the extent of deafness.

Atypical Cogan syndrome2 occurs when types of ocular inflammation other than interstitial keratitis happens in conjunction with hearing loss. We describe 3 adolescents with atypical Cogan syndrome3 who lost all of their hearing owing to sensorineural involvement.

Report of Cases. Case 1. A 16-year-old African American adolescent boy was seen 4 months after an acute loss of all of his hearing, which developed during 1 week. For the past 4 months, his eyes were constantly red. His pediatrician had prescribed him systemic antibiotics to be taken orally. Visual acuity was 20/25 OU with an intraocular pressure of 12 mm Hg OU. He had mild cells and flare in each eye with 360° of posterior synechiae in each eye.

Figure 6. Diffuse positive immunoreactivity to CD34 (immunohistochemical stain, original magnification ×100).