ventricular wall (Figure 2A). Color flow imaging demonstrated blood flow within the deep recess between the trabeculations (Figure 2B). Severe mitral regurgitation due to grade 1 anterior mitral valve prolapse was also present. Findings on the carotid Doppler study were normal, and no signs of carotid stenosis or plaques were observed.

A provisional diagnosis of noncompaction cardiomyopathy was made. Cardiac magnetic resonance imaging was recommended to further evaluate the cardiomyopathy, but the patient declined. Long-term oral warfarin sodium treatment was commenced to reduce the risk of systemic embolization. At 1 month after the attack, his visual acuity remained similar in the right eye and improved to 20/200 OS.

Comment. The embryonic arrest of compaction of myocardial fibers seen in noncompaction cardiomyopathy is most frequently observed in the left ventricle. The cardiomyopathy is diagnosed by echocardiography or magnetic resonance imaging.

Echocardiography shows trabeculations and deep intertrabecular recesses. Blood flow can be observed within the deep intertrabecular recesses, and the flow is in continuity with the left ventricular cavity. Noncompaction cardiomyopathy is diagnosed echocardiographically when the ratio of trabeculations to the thickness of the underlying ventricular wall is more than 2.

Magnetic resonance imaging shows a 2-layered wall structure comprising a thin compacted epicardium and a thick noncompacted myocardium. Our patient’s echocardiograms are consistent with the diagnosis of noncompaction cardiomyopathy.

Strokes have been reported as a systemic thromboembolism that occurs in patients with noncompaction cardiomyopathy. The bilateral retinal artery occlusion seen in our patient is likely of a thromboembolic nature. We postulated that the microembolus observed in the left retinal arteriole originated from the heart. The noncompaction cardiomyopathy with relative blood stasis in the intertrabecular recess explains the most probable cause of this phenomenon. Thus, it is extremely important to highlight this rare cause of retinal artery occlusion that has resulted in devastating vision loss.

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Conflict of Interest Disclosures: None reported.

Multiply Recurrent Solitary Fibrous Tumor of the Orbit Without Malignant Degeneration: A 45-Year Clinicopathologic Case Study

Solitary fibrous tumor (SFT) is a rare mesenchymal spindle cell neoplasm originally described in the pleura and subsequently identified in a number of extrathoracic sites. Orbital SFT was first de-
scribed in 1994; since then, more than 100 cases have been reported or reclassified with that diagnosis.1,2 Only 4 patients were younger than 10 years when first diagnosed as having orbital SFT. While most reports describe a benign clinical course, rare cases of primary malignant orbital SFT have also been documented. This led the World Health Organization to classify SFT as a neoplasm of intermediate biological potential (locally aggressive, rarely metastasizing).3,4 Increased cellular atypia or mitotic activity in recurrent lesions following primary excision of orbital SFTs has also been described, suggesting that complete initial resection is a critical prognostic factor in preventing malignant degeneration.1,4-7

We describe a patient with orbital SFT whose proptosis was first recognized at age 9 years and who underwent surgical excision at various institutions at ages 12, 22, and 52 years. To our knowledge, this represents the longest histopathologically documented follow-up of a patient with orbital SFT.

Report of a Case. In 1967, a 9-year-old girl with painless left proptosis underwent an exploratory craniotomy in which no tumor was found. With a presumptive diagnosis of orbital hemangioma, she was treated with external beam radiation. In 1970, she underwent an orbitotomy with removal of a 4.0 × 2.5-cm lobulated, red-dish-blue mass, described as a hemangioma. Recurrent proptosis was noted in 1980, and a lateral orbitotomy with bone flap was performed with removal of a 5-cm lesion. At that time, review at the Armed Forces Institute of Pathology of the 1970 and 1980 specimens yielded a diagnosis of fibrous histiocytoma.

Figure 1. Axial (A) and coronal (B) computed tomographic images demonstrating a left 2.7×1.7×1.6-cm mass isodense to muscle (A) and with homogeneous intense contrast enhancement (B).

Figure 2. Gross tumor following excision.

Figure 3. Short intersecting fascicles of cytologically bland spindle cells arranged in a “patternless pattern” (hematoxylin-eosin) (A), dilated hemangiopericytic thin-walled vessels (hematoxylin-eosin) (B), and positive CD34 immunostaining (C) (original magnification ×200).
In 2010, the patient was referred to our institution with a history of gradually recurrent proptosis and recent-onset diplopia. Orbital imaging demonstrated a large, homogeneously enhancing mass that filled the inferotemporal extraconal and intracranal left orbit (Figure 1). A lower fornix-approach orbitotomy was performed with removal of all gross tumor (Figure 2). Histologic sections showed a proliferation of spindle cells with eosinophilic cytoplasm, poorly defined cytoplasmic borders, and elongated nuclei with small eosinophilic nucleoli, arranged in short intersecting fascicles in a “patternless pattern” (Figure 3A). Occasional hemangiopericytic thin-walled vessels were present (Figure 3B). Rare mitotic figures were seen, with no atypical forms, areas of necrosis, or cytologic atypia identified. Immunohistochemical staining revealed that neoplastic cells were strongly and diffusely positive for CD34 (Figure 3C), CD99, and Bcl-2 and negative for smooth muscle actin, muscle-specific actin, desmin, S-100 protein, and AE1/AE3 cytokeratins, supporting the diagnosis of SFT. The patient did well following gross tumor excision and remained symptom free at 1 year.

Comment. We are aware of 4 cases of recurrent SFT that showed increased cellularity, atypia, or mitotic rate compared with the primary lesions, suggesting malignant transformation with time.6,7 Such cases have led to the conclusion that the most important prognostic factor is not the initial histologic appearance but rather complete primary resection.1 In those reports, the interval between initial tumor excision and final recurrent tumor excision ranged from 6 months to 7 years. Our case provides further insight into the heterogeneous histologic and clinical behavior of these tumors. After an interval that included initial recognition of proptosis at age 9 years and multiple surgical resections as late as age 52 years, there were no histopathologic features to suggest malignant transformation. A limitation of this study is that the 1970 and 1980 outside histopathologic material is no longer available to document a uniform diagnosis throughout the 45-year course. However, a 1980 Armed Forces Institute of Pathology review of that material did yield a diagnosis of orbital fibrous histiocytoma, an entity that has been more recently reclassified as orbital SFT in many cases.2 Our case suggests that incompletely excised, long-standing SFTs do not necessarily increase in biological aggressiveness.

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Author Contributions: Drs Griepentrog and Harris had full access to all the data in the study and take responsibility for the integrity of the data and the accuracy of the data analysis.

Conflict of Interest Disclosures: None reported.

Funding/Support: This work was supported in part by an unrestricted grant from Research to Prevent Blindness.


COMMENTS AND OPINIONS

ω-3 Intake in Patients With Retinitis Pigmentosa Receiving Vitamin A

Berson and colleagues1 claim that visual acuity loss is slower in adults with retinitis pigmentosa receiving vitamin A who also consume a diet rich in ω-3 fatty acids. They previously reported findings of a clinical trial of nutritional supplementation for patients with retinitis pigmentosa.2 They were criticized in 2 subsequent publications3,4 for overstating the strength of evidence for their clinical recommendations. The same can be said for this article.

A suggestive fragment of data taken from an epidemiologic study, not a randomized trial, is offered as a basis for clinical therapy. No biological mechanism is offered as motivation or interpretation of this research. The groups compared are self-selected in their dietary choices, not randomly assigned. Such studies are ordinarily thought of as starting points for clinical research. If other nutritional factors in addition to ω-3 intake were evaluated for association with visual acuity changes, tests of significance should be modified.

The authors point out that the cut points for high and low ω-3 fatty acid intake were taken from a previous trial with different treatments and visual outcomes, and they reassure us that the high- and low-intake groups in this article are balanced. No such assurance is offered for the median and quartile data. A regression analysis to see whether a trend exists would be helpful.

Ophthalmologists need to be cautioned not to allow this article to influence their management of patients with retinitis pigmentosa, despite the strong conclusions offered by these investigators.

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