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SHORT COMMUNICATION

Bilateral atypical nodular posterior scleritis

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PURPOSE. *To evaluate ocular features of nodular posterior scleritis simulating choroidal melanoma.*
METHODS. *A 60-year old woman presented with blurred vision in her right eye of two weeks duration. On examination she had a mild right-globe proptosis with an episcleral nodular mass as well as a large elevated nonpigmented choroidal mass involving the nasal quadrant.*
RESULTS. *A and B-scan ultrasonography showed a medium to high-reflective solid choroidal mass. MRI demonstrated a bi-convex mass in the medial aspect of the right globe with signal characteristics compatible with choroidal melanoma. Biopsy of the extra ocular lesion demonstrated chronic inflammatory cell infiltrate suggestive of posterior scleritis. She responded to corticosteroid therapy. On evaluation 41 months later she was noted to have a similar choroidal mass in the left eye.*
CONCLUSIONS. *The physician should be aware of the clinical manifestations and diagnostic hallmarks of nodular posterior scleritis in order to differentiate this inflammatory process from choroidal melanoma. (Eur J Ophthalmol 2006; 16: 614-7)*

KEY WORDS. *Nodular, Posterior scleritis, Choroidal melanoma*

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INTRODUCTION

Nodular posterior scleritis typically presents as a solid subretinal mass and can be mistaken for a choroidal malignant melanoma (1-9). Subretinal lesions manifest by posterior scleritis can also resemble carcinoma metastatic to the uvea, circumscribed choroidal hemangioma, or benign reactive uveal lymphoid hyperplasia (1).

We describe an atypical bilateral nodular posterior scleritis with associated orbital pseudotumor that simulated a choroidal melanoma with extraocular extension.

Case report

In September 1994, a 60-year-old woman presented with a chief complaint of blurred vision in the right eye of 2 weeks duration. She also described mild discomfort and a sense of bulging of her right eye.

On examination, visual acuity was correctable to 20/40 in the right eye and 20/20 in the left. External examination revealed mild right globe proptosis. Slit lamp examination showed two episcleral or scleral nodular masses on the supranasal aspect of the globe. Ophthalmoscopy of the right eye revealed a prominent elevated nonpigmented subretinal mass involving the nasal quadrant and extending from approximately the 2 o'clock to the 5 o'clock meridian clockwise and an overlying serous detachment of the neurosensory retina (Fig. 1). Examination of the left eye revealed no pertinent abnormalities.

Fluorescein angiography documented the subretinal mass but did not reveal any large caliber intralesional blood vessels or significant leakage (Fig. 2). B-scan ultrasonography showed a sausage-shaped thickening of the eye wall tissues nasally, surrounded by a retrobulbar echolucent zone. A-scan revealed moderately high amplitude reflectivity of the entire mass (Fig. 3). Orbital magnetic resonance imaging (MRI) demonstrated a biconvex mass involving the medial wall of the right eye measuring 2 x 1 x 2.5 cm as well an orbital soft tissue mass interpreted as extrascleral extension of the choroidal mass in

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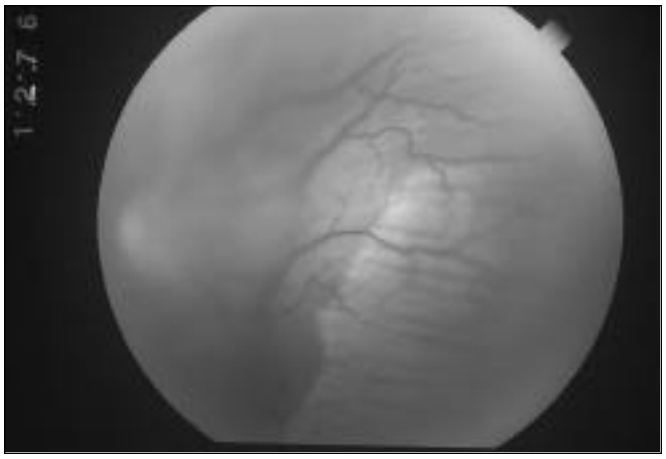

Fig. 1 - *Fundus photograph of the right eye shows a prominent non-pigmented subretinal mass involving the nasal quadrant.*

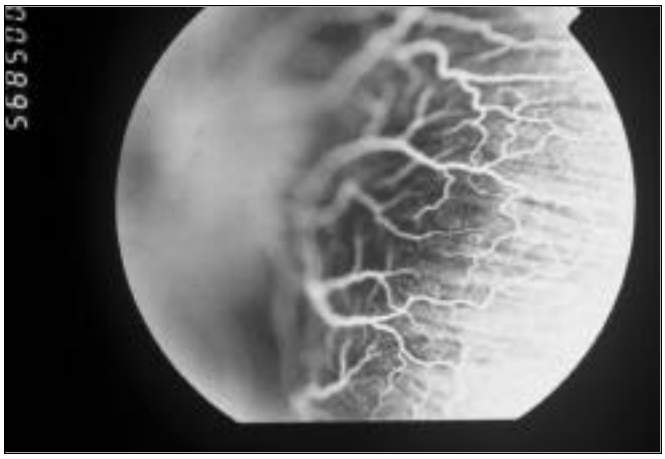

Fig. 2 - *Fluorescein angiography documented the subretinal mass without any large caliber intralesional blood vessels or significant leakage.*

the inferonasal aspect of the orbit measuring 1.5 x 0.8 x 0.1 cm. The mass was of intermediate signal intensity on the T1-weighted images and hypointense on the T2-weighted images. It exhibited fairly homogenous gadolinium enhancement in the periphery and less centrally (Fig. 4). The MRI findings were interpreted by the radiologists as consistent with choroidal melanoma with extrascleral extension.

In view of the MRI findings and radiologic interpretation, biopsy of the extraocular lesion was performed. Two sections of a firm white mass adherent to approximately 5 to 6 mm from the surgical limbus were excised. The globe specimens revealed scleral tissue that was moderately infiltrated with chronic inflammatory cells and lacked the normal orderly arrangements and compactness. Some sections showed scleral necrosis along with increased inflammation. No tumor cells were identified. Based on the histopathologic and ultrasonographic findings, our presumptive post-biopsy diagnosis was nodular posterior scleritis associated with localized orbital pseudotumor.

The patient was treated with an intense 3-day course of intravenous corticosteroids. Within 72 hours her fundus mass was distinctively flatter and her proptosis in the right eye decreased. Her vision improved to 20/15 and her ocular discomfort diminished to a dull ache.

Following the initial treatment, the patient was continued on tapering oral prednisone for the next 7 months. Her ocular pain diminished, the ocular redness decreased slightly, the fundus mass became less prominent, and the proptosis was gone. At follow-up evaluation 41 months

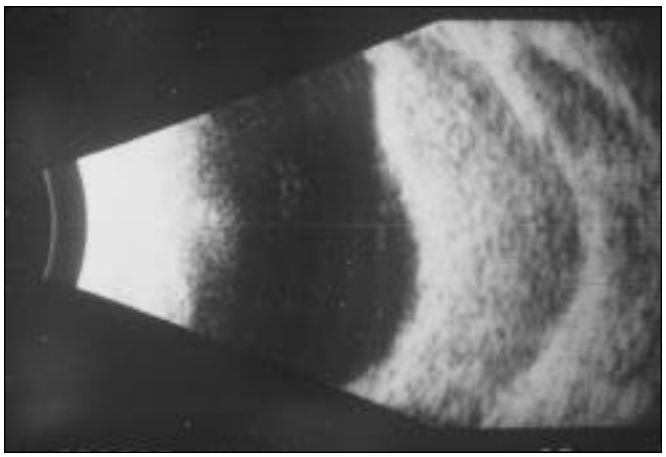

Fig. 3 - *B-scan ultrasonography shows a prominent sausage-shaped mass, surrounded by a retrobulbar echolucent cleft.*

after her initial treatment, a new fundus mass was noted in the left eye. The mass was similar to the previously noted mass in the right eye. It was also located in the inferonasal midzone. The new mass did not respond to periocular injections of corticosteroids.

Over the next several years, the ridge-like eye wall mass bilaterally extended slowly in a circumferential direction resulting in a fundus appearance resembling the indentation caused by a high encircling scleral buckle.

About 5 years after the initial treatment, the patient began to experience diplopia on extreme right gaze. She was found to have restriction of abduction in the right eye due to epibulbar fibrosis around the medial rectus muscle insertion. Additional eye wall tissue was excised medially in the right eye in an attempt to eliminate the restriction of

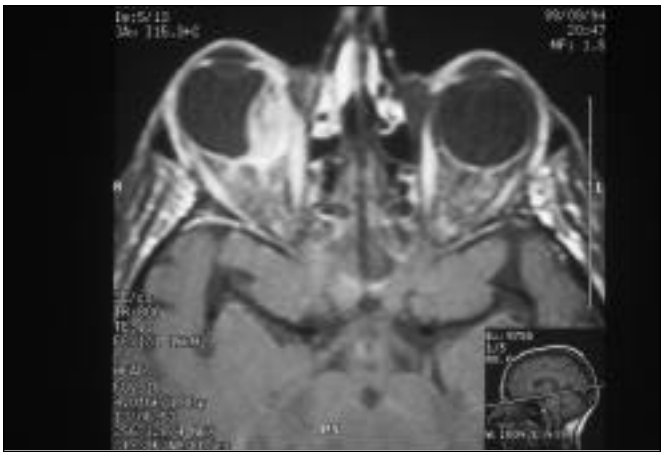

Fig. 4 - *Magnetic resonance imaging reveals a biconvex mass involving the medial wall of the right eye.*

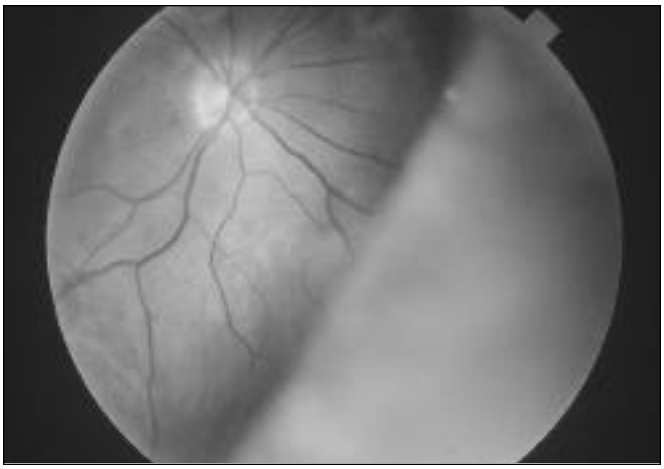

Fig. 5 - *Fundus photograph of the right eye taken 9 years later reveals an encircling ridge-like mass, resembling encircling scleral buckle.*

abduction and provide supplemental tissue specimen for pathologic analysis. Pathologic study of this tissue showed disorganized sclera containing numerous chronic inflammatory cells and scattered foci of granulomatous inflammation without caseation or geminal centers.

At most recent ophthalmic follow-up examination 9 years following initial treatment, visual acuity was 20/40 bilaterally (Fig. 5). The retina remained fully attached in each eye.

DISCUSSION

Nodular posterior scleritis is the rarest form of scleritis and can be clinically perplexing in the absence of classic symptoms such as ocular pain and ocular redness (2, 3, 5-7). Unilateral in predilection for the female sex, posterior scleritis is often associated with anterior scleritis (4). The posterior inflammatory process often precedes anterior inflammation. Funduscopically, the appearance of nodular posterior scleritis can resemble choroidal melanoma, choroidal metastasis, or benign reactive uveal lymphoid hyperplasia. In a comprehensive review of 400 patients with lesions clinically simulating choroidal melanoma, nodular posterior scleritis was documented in 6 of the 400 patients (1.5%) (5).

Several features can help to distinguish posterior scleritis from a neoplasm. Patients with posterior scleritis have pain and anterior scleritis (1, 8), although these are not universally present. In addition, the fundus mass is characteristically the same orange color as the adjacent uninvolved fundus, has a normal choroidal vascular pattern, and is typically surrounded by choroidal folds or retinal striae (1, 8).

Our patient presented with symptoms suggestive of orbital pseudotumor. When the eye on the affected side was examined, an extensive, nonpigmented, subretinal mass and associated serous detachment of the neurosensory retina was observed. Fluorescein angiography was not helpful diagnostically.

Orbital MRI was grossly misinterpreted. The ancillary test that proved most informative was ultrasonography. It confirmed other authors' findings of a sausage-shaped thickening of the wall with high internal reflectivity and retrobulbar echolucency (6, 7). These findings coupled with the biopsy findings of inflammatory cells with a disorganized and partially necrotic sclera led to the correct diagnosis of nodular posterior scleritis. Interestingly, 5 years after the original presentation of a choroidal mass in the right eye, the patient developed a similar eye wall mass in the inferonasal midzone in the contralateral eye.

To our knowledge, this has not been previously described in the literature. Our case is also unique in that the ridge-like eye wall mass extended slowly in a circumferential direction resulting in a fundus appearance resembling the indentation caused by a high encircling scleral buckle. In spite of advances in imaging techniques, diagnosis of nodular scleritis can still present a diagnostic challenge.

No authors have any proprietary interest.

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REFERENCES

1. Brod RD, Saul RF. Nodular posterior scleritis. Arch Ophthalmol 1990; 108: 1170-1.
2. Benson WE. Posterior scleritis. Surv Ophthalmol 1988; 32: 297-316.
3. Watson PG, Hayreh SS. Scleritis and episcleritis. Br J Ophthalmol 1976; 60: 163-91.
4. Portnoy SL, Franklin RM. Retina, Second Edition, Vol. 2. St. Louis: CV Mosby, 1999; 1651-9.
5. Shields JA, Augsburger JJ, Brown GC, Stephens RE. The differential diagnosis of posterior uveal melanoma. Ophthalmology 1980; 87: 518-22.
6. Finger PT, Perry HD, Packer S, Erdrey RA, Weisman GD, Sibony PA. Posterior scleritis as an intraocular tumor. Br J Ophthalmol 1990; 74: 121-2.
7. Demirci H, Shields CL, Honavar SG, Shields JA, Bardebstein DS. Long term follow up of giant nodular posterior scleritis simulating choroidal melanoma. Arch Ophthalmol 2000; 118: 1290-2.
8. Feldon SE, Sigelman J, Albert DM, Smith TR. Clinical manifestations of brawny scleritis. Am J Ophthalmol 1978; 85: 781-7.
9. Yap EY, Robertson DM, Buettner H. Scleritis as an initial manifestation of choroidal melanoma. Ophthalmology 1992; 99: 1693-7.