Long-term Follow-up of Giant Nodular Posterior Scleritis Simulating Choroidal Melanoma

Hakan Demirci, MD; Carol L. Shields, MD; Santosh G. Honavar, MD; Jerry A. Shields, MD; David S. Bardenstein, MD

A 41-year-old asymptomatic woman was referred for enucleation of a 7.5-mm-thick intraocular tumor suspected to be choroidal melanoma. The clinical findings combined with imaging studies suggested instead a diagnosis of giant nodular posterior scleritis. A scleral biopsy was performed to confirm the diagnosis. After 12 years of observation, the lesion has remained stable and visual acuity has been preserved. Nodular posterior scleritis can present with no symptoms of pain, redness, or visual disturbance and can remain quiet for many years. It must be clinically differentiated from choroidal melanoma.

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Several benign and malignant fundus lesions can clinically resemble choroidal melanoma and pose a diagnostic challenge.1 In a large review of 400 consecutive patients with lesions clinically simulating choroidal melanoma, the most common pseudomelanomas were suspicious choroidal nevus (27%), age-related macular degeneration (13%), age-related extramacular degeneration (11%), congenital hypertrophy of the retinal pigment epithelium (10%), choroidal hemangioma (8%), and nodular scleritis (1.5%).1 Nodular posterior scleritis is uncommon and rarely does it present with such dimensions as to simulate a choroidal melanoma. However, failure to recognize it could lead to misdirected treatment.2

We present a 12-year follow-up of a case of nodular posterior scleritis that closely simulated a choroidal melanoma and in which the diagnosis was established based on the clinical findings, imaging techniques, and histopathologic examination.

REPORT OF A CASE

On routine examination in December 1987, a 41-year-old asymptomatic woman was found to have an amelanotic mass in the fundus of the left eye. The patient was referred to the Ocular Oncology Service at Wills Eye Hospital (Philadelphia, Pa) for possible enucleation of a suspected choroidal melanoma. Her visual acuity was 20/20 OU and intraocular pressures were normal. There were no cells in the anterior chamber, synechiae, or pupillary abnormalities in either eye. The right eye was normal.

In the left eye, an atypical, hyperemic pterygium encroached over the nasal cornea. There was no scleral necrosis or ectasia. On fundus examination, there was a dome-shaped subretinal mass measuring 13 × 12 mm basally and 7.5 mm in thickness, located in the inferonasal quadrant with slight serous subretinal fluid overlying it (Figure 1). There were no choroidal folds or vitreous cells. The lesion was slightly darker orange than the adjacent normal choroid and had a subtle darker circumferential base. The slightly tortuous retinal blood vessels and normal choroidal vasculature, however, were seen on the inner surface of the lesion (Figure 2). On transcleral transillumination, the lesion transmitted light readily, suggestive of a nonmelanocytic process.

On intravenous fluorescein angiography, relative hypofluorescence of the mass with the hyperfluorescence of the abnormal retinal vessels was noted in the early phases and stayed hypofluorescent in late phases (Figure 3). There was no

From the Oncology Service, Wills Eye Hospital, Thomas Jefferson University, Philadelphia, Pa (Drs Demirci, C. Shields, Honavar, and J. Shields); and the Department of Ophthalmology, Case Western University, Cleveland, Ohio (Dr Bardenstein).
“double circulation” or scleral staining. On A-scan ultrasonography, high-amplitude internal reflectivity was observed. B-scan ultrasonography showed a prominent, dome-shaped mass at the sclerochoroidal level with acoustic solidity and shallow retrobulbar echolucent cleft (Figure 4). Magnetic resonance imaging showed a low-signal scleral mass with slight gadolinium enhancement along the internal margin of the lesion (Figure 5).

The clinical findings of orange tumor color, apparently normal choroidal vasculature pattern on inner-tumor surface, light transmission on transillumination, hypofluorescence without double circulation on intravenous angiography, and mass with acoustic solidity associated with a shallow retrobulbar echolucent cleft on B-scan ultrasonography were all suggestive of posterior scleritis rather than choroidal melanoma. Because of the patient’s anxiety and referring physician’s concern, a scleral trephine punch biopsy was performed. A posteriorly hinged scleral flap of 0.8 mm in thickness and 8 mm in diameter was developed over the mass. A 2.5-mm trephine was
placed to a depth of 7 mm in the scleral bed, carefully watching for uveal tissue. After excision of the biopsy material, the scleral flap was sutured back into position. Histopathologic examination of the biopsy material showed foci of chronic inflammatory cells (lymphocytes and eosinophils) within the thickened scleral collagen, and increased fibroblasts compatible with scleritis (Figure 6). Systemic evaluation revealed no collagen vascular disease.

The patient remained asymptomatic and was examined yearly. After a follow-up of 12 years, her visual acuity remained 20/20 OU. The subtle conjunctival injection over the surface of nasal conjunctiva persisted and the fundus lesion remained unchanged.

COMMENT

During the past decades, clinicians have become more experienced and accurate in the diagnosis of choroidal melanoma. Of more than 12,000 patients with uveal melanoma managed on the Ocular Oncology Service at Wills Eye Hospital, a less than 1% misdiagnosis rate was noted. This demonstrates great improvement on previous misdiagnosis rates of nearly 20% as reported by the Armed Forces Institute of Pathology (Washington, DC). Despite growing experience and diagnostic advances, some simulating choroidal lesions continue to be a diagnostic challenge. Nodular posterior scleritis is known to simulate malignant melanoma. Of 400 cases referred to the Ocular Oncology Service diagnosed as choroidal melanoma, 6 (1.5%) proved clinically to be posterior scleritis. In the literature, there are reports in which clinical misinterpretation of posterior scleritis as choroidal melanoma has led to therapeutic misdirection, including enucleation. It is important to recognize the distinguishing features between posterior scleritis and choroidal melanoma. In a report on 137 patients with posterior scleritis, McCluskey and coworkers and Calthorpe et al found that 65% of them were female and 29% had an associated systemic condition. The most common symptoms were those associated with anterior scleritis (60%), pain (56%), and poor vision (31%). The most common signs were serous retinal detachment (21%), optic nerve swelling (18%), and circumscribed fundus mass (13%). It is noteworthy that our patient had no symptoms or associated collagen vascular disease. The only finding was nonpigmented fundus mass with minimal subretinal fluid. We believe that the nonpigmented appearance of the lesion, normal intrinsic choroidal vascular pattern, and transmission of light on transillumination were important clinical clues suggestive of posterior scleritis.

Ancillary tests can be helpful in the diagnosis of posterior scleritis. Ultrasonography generally reveals an echogenic mass with echo-free retrolubular edema. Additionally, magnetic resonance imaging can be helpful with tumors larger than 2 mm thick, and usually shows a low signal relative to vitreous in both T1- and T2-weighted images with variable enhancement with gadolinium. In equivocal cases, biopsy can be revealing but it should be understood that biopsy of a transcleral mass can be hazardous and it should be reserved for selected diagnostic problems.

Systemic, retrobulbar, or topical corticosteroids are generally recommended for symptomatic posterior scleritis. The natural history of posterior scleritis is characterized by intermittent pain, swelling, and even scleral necrosis, but our case is unique in that the massive lesion has remained stable during 12 years of observation. Despite growing clinical experience and advances in imaging techniques, differentiation between posterior scleritis and malignant melanoma can be a challenge.

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Reprints: Carol L. Shields, MD, Ocular Oncology Service, Wills Eye Hospital, 900 Walnut St, Philadelphia, PA 19107.

REFERENCES