Giant Choroidal Nevus Followed for 22 Years

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We introduce to you a new section in Retina Today entitled Case Reports in Ocular Oncology. The purpose of this section is to provide the retina specialist with a potpourri of instructive cases, thoughts, short reviews, or new techniques in ocular oncology in both adults and children. The column will be well illustrated with one or several imaging techniques to depict the relevant features of the described lesion. Important points relative to tumor identification, diagnosis, and therapy will be discussed. We will attempt to focus on

the newest developments in ocular oncology, and we hope to provide education on some controversial issues.

In this installment, the clinical features of choroidal nevus are discussed, and the authors describe a rare variant of nevus, the giant choroidal nevus (over 10 mm in diameter). This is often mistaken for melanoma and can pose a diagnostic dilemma. The authors mention the classic features of choroidal nevi and then explain the features of giant choroidal nevi with a focus on the ultimate long-term outcomes of these tumors.

We extend an invitation to readers to submit informative cases, thoughts, short reviews, or new techniques to Retina Today. Please send submission for consideration to Rachel Renshaw at rrenshaw@bmctoday.com. We look forward to this exciting new column, and we guarantee that it will be filled with color.

-Carol L. Shields, MD; and Sara Lally, MD The Ocular Oncology Service, Wills Eye Institute

46-year-old white male was found on routine examination to have a large pigmented lesion in the fundus of his right eye. The patient had no personal or family history of ocular trauma, disease, or cancer.

On examination, visual acuity was 20/20 in the right eye and 20/15 in the left eye. Slit-lamp examination and intraocular pressures were normal in each eye. Fundus examination of the left eye was normal. Fundus examination of the right eye disclosed a pigmented lesion occupying the nasal region and measuring 15 x 12 mm in basal dimensions and 2.2 mm in thickness. The lesion was 1 mm nasal to the disc. Trace chronic subretinal fluid, retinal pigment epithelial (RPE) atrophy and hyperplasia, and numerous drusen were found overlying the lesion with areas of fibrous metaplasia of the RPE. The patient was diagnosed with giant choroidal nevus vs dormant choroidal melanoma.

Observation twice yearly with photographic documentation was provided. Twenty-two years later the giant choroidal nevus remained stable with no signs of growth, progressive subretinal fluid, or orange pigmentation. Over time, the picture evolved slightly, with change in drusen appearance and increased overlying RPE atrophy (Figure 1). Currently, the tumor thickness remains



Figure 1. Fundus photo of a stable pigmented choroidal lesion over 22 years measuring 15x12x2.2 mm with extensive overlying RPE atrophy elasia and numerous drusen.

stable with features of acoustic solidity and no subretinal fluid (Figure 2). The patient continues to be followed with a stable giant choroidal nevus.

DISCUSSION

Choroidal nevus is a benign melanocytic lesion, appear-

Feature	P value	Confidence interval (95%)	Relative risk
Nevus proximity to foveola mm (mean)	0.049	(1.00-1.29)	1.14 ^a
Nevus proximity to foveola (0 mm vs 0 mm)	0.037	(1.07-7.11)	2.75
Tumor thickness mm (mean)	0.033	(1.06-3.63)	1.96 ^b
Acoustic quality (hollow vs not hollow)	0.048	(1.01-5.45)	2.34

ing pigmented or non-pigmented, and classically less than 2 mm in thickness.¹ Although uncommon, malignant transformation of choroidal nevi into melanoma is possible.^{1,2} It has been estimated that one in 8,845 choroidal nevi transform into melanoma in the white population.³ The Blue Mountains Eye Study surveyed a large population-based sample in Australia and found that nevi were present in 6.5% of the general population over age 49, with a slight decrease in prevalence with increasing age.⁴ Shields and associates reported that factors predictive of growth into melanoma include tumor thickness greater than 2 mm, subretinal fluid, symptoms, orange pigment, tumor margin within 3 mm of the optic disc, ultrasonographic hollowness, and halo absence.⁵

Nevi are generally small, with a mean nevus diameter of 1.25 mm according to the population-based Blue Mountains Eye Study,⁴ but a clinic-based study from Wills Eye Institute Oncology Service found mean diameter of choroidal nevus to be larger at 5.1 mm.⁵ This discrepancy is likely related to referral bias of more suspicious nevi to an oncology service. Choroidal nevi rarely affect visual acuity unless they are located in a subfoveal site.⁶ According to the Blue Mountain Eye Study,⁴ there are no significant associations between choroidal nevus and iris or skin color or sun-induced skin damage.

Large choroidal nevi over 10 mm in diameter are extremely rare and are classified as giant choroidal nevi. Li and associates studied 322 patients with giant nevi (diameter greater than 10 mm) and found that these lesions commonly simulate melanoma.² However, there are features that suggest a benign lesion, such as the presence of drusen (81%), RPE atrophy (20%), RPE hyperplasia (15%), and fibrous metaplasia (15%).² Additionally, giant nevi tend to lack acoustic hollowness on ultrasound, orange pigmentation, and serous retinal detachment. 2 In that series, 13% grew into melanoma by 5 years and 24% by 15 years.² Most proved to be low-grade melanoma with extremely slow growth and minimal subretinal fluid. Features predictive of growth included acoustic hollowness and close proximity to the foveola (Table).²

In summary, most choroidal nevi are relatively small lesions of 5 mm diameter. Occasionally choroidal nevi,

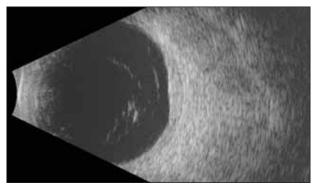


Figure 2. B-scan ultrasound showing the nevus as an acoustic solid choroidal mass and without subretinal fluid.

however, are giant and measure over 10 mm diameter. Long-term follow-up of these patients is crucial to monitor for transformation into melanoma. Our patient showed no change over 22 years.

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