RPE Detachment Overlying Choroidal Nevus Imparting Pseudo-growth Appearance

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Choroidal nevus is the most common primary intraocular tumor; it classically presents as a pigmented lesion with overlying drusen and retinal pigment epithelial (RPE) alterations.[1] Visual acuity in patients with choroidal nevi is mostly unaffected but can decline over time when associated with subfoveolar location of the tumor, orange pigment, subretinal fluid or RPE detachment (RPED).[2] We describe here a case of choroidal nevus with an overlying RPED and discuss its clinical and prognostic implications.

CASE DESCRIPTION

A 53-year-old asymptomatic white man was referred for evaluation of a pigmented fundus lesion in his left eye (OS) discovered during routine examination.

On initial presentation, visual acuity was 20/20 in each eye. Slit-lamp examination and intraocular pressures were within normal limits in both eyes. Dilated fundus examination of the right eye was unremarkable. The left eye disclosed a pigmented choroidal lesion inferior to the optic disc and measuring 3.5 x 3.5 mm in basal dimensions and 1 mm in ultrasonographic thickness. Soft drusen were present over the lesion. The mass was acoustically solid on B-scan ultrasonography. Orange pigment, subretinal fluid, and RPE alterations were absent. The clinical findings were consistent with choroidal nevus, and observation was recommended.

After 3 years of stable follow-up, the mass increased slightly in thickness ultrasonographically. Clinically, a transparent overlying RPED had formed, evident on autofluorescence, and more clearly visible on fluorescein angiography as mottled, well-demarcated hyperfluorescence with late pooling, and on optical coherence tomography (OCT) as an abrupt dome-shaped elevation at the level of the RPE (Figure 1). There was no hemorrhage, exudation, subretinal fluid, or sign of choroidal neovascularization. Observation was advised, and the RPED remained stable over a period of 9 years with visual acuity remaining 20/20 in both eyes.

DISCUSSION

RPED is a somewhat uncommon and under-reported finding associated with choroidal nevus. In the population-based Blue Mountain Eye Study, choroidal nevi were found in 6.5% of the 3,654 patients (age 49 to 97 years), but RPED was not described as an associated feature.[3] In clinic-based reports on 3,422 and 120 eyes with choroidal nevi monitored at Wills Eye Institute, RPED was detected in 1.2% of cases clinically and in 12% of cases by OCT.[1,4] In a separate analysis, RPED was not a risk factor for growth or transformation of nevus to malignant melanoma but was found to be a predictor of poor visual outcome, particularly for subfoveal nevi.[2,5] In an analysis of visual outcomes in 3,422 patients with choroidal

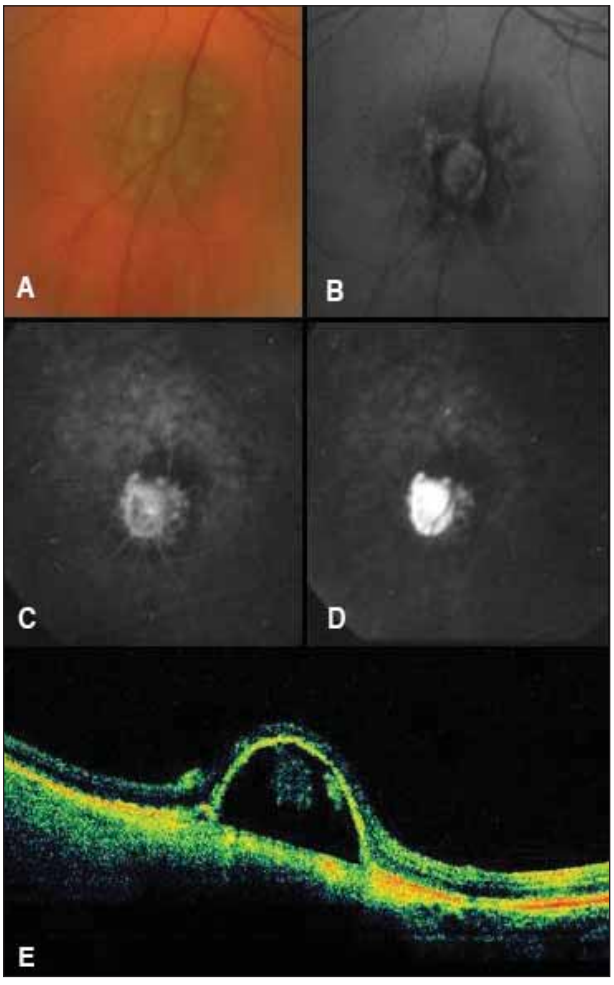

Figure 1. Choroidal nevus with fine overlying drusen in the periphery and subtle serous RPE detachment seen after 3 years of stable follow-up (A). The RPED is hyperautofluorescent (B) and shows early mottled hyperfluorescence (C) and late circumscribed pooling (D) on fluorescein angiography. Optical coherence tomography (E) clearly defines the elevation at the level of the RPE.

nevi, RPED, particularly in the fovea, carried a relative risk of 22.2 for loss of 3 or more logMAR lines of vision and a relative risk of 12.8 for final visual acuity of less than 20/200.[2] Often undiagnosed by clinical exam alone, RPED can be better detected with OCT, allowing confirmation of the diagnosis and better prognostic information for visual outcome.

Histopathologically, the RPE is juxtaposed between the outer segment of the photoreceptor layer and Bruch membrane.[6] Its functions include formation of the outer blood-retinal barrier, phagocytosis of photoreceptor outer segments, retinol metabolism, and transport of nutrients and metabolic end products of the retina, all of which help to maintain the integrity of the retina.[6] An RPED overlying a choroidal nevus likely occurs secondary to degenerative changes in Bruch membrane.[6] With time, there is an increased amount of lipid and waste products deposited in Bruch membrane leading to hydrophobic barrier formation and the accumulation of sub-RPE fluid.[6-9]

RPED are classified etiologically (inflammatory, ischemic, idiopathic, and degenerative) or clinically (serous, drusenoid, and fibrovascular).[6] Our case demonstrates a serous RPED, described as a smooth, sharply demarcated, dome-shaped RPE elevation that contains predominantly serous fluid.[6] This can best be visualized as an abrupt, clear elevation on OCT and also demonstrated on fluorescein angiography with early hyperfluorescence and well-circumscribed late pooling.[6] Autofluorescence demonstrates moderate localized hyperautofluorescence that is particularly bright at the margins of the RPED.[6,7] An analysis of autofluorescence of choroidal nevi in 64 patients revealed RPED hyperautofluorescence when the detachments appeared fresh and hypoautofluorescence in RPED with chronicity.[8]

While RPEDs overlying choroidal nevi often contain serous fluid alone, investigation for choroidal neovascular membrane (CNVM) should be made, as this finding can adversely affect visual prognosis.[6] Diagnostic clues indicating presence of CNVM in RPED include associated serous retinal detachment, presence of exudation and/or blood, and chorioretinal folds.[6] Fluorescein angiography, indocyanine green angiography, and OCT can help visualize occult CNVM within RPED.[6,10]

Treatment of CNVM secondary to choroidal nevus varies. Levy and associates reported a case of a 52-year-old man with a fibrovascular RPED extending into the fovea secondary to a choroidal nevus, which was treated successfully with two sessions of photodynamic therapy with improvement of visual acuity from 20/120 to 20/40.[11] Other treatment modalities include laser photocoagulation, transpupillary thermotherapy, and vascular endothelial growth factor inhibitors.[12-14]

SUMMARY

In summary, RPED overlying choroidal nevus is a somewhat uncommon clinical finding that can be confirmed with ancillary testing. This is important because an overlying RPED can pseudo-enlarge a choroidal nevus, causing suspicion for transformation into melanoma. RPED is not associated with malignant transformation, but on the contrary implies stability and chronicity of the nevus. RPED overlying a subfoveal nevus is a long-term risk for visual loss; therefore, careful clinical examination and

appropriate diagnostic procedures should be implemented for all patients with choroidal nevi. ■

Support provided by the Eye Tumor Research Foundation, Philadelphia, PA (CLS). The funders had no role in the design and conduct of the study; in the collection, analysis, and interpretation of the data; and in the preparation, review or approval of the manuscript. Carol L. Shields, M.D. has had full access to all the data in the study and takes responsibility for the integrity of the data and the accuracy of the data analysis.

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The authors have no financial interests in the products mentioned in this case report.

1. Shields CL, Furuta M, Mashayekhi A, et al. Clinical spectrum of choroidal nevi based on age at presentation in 3422 consecutive eyes. *Ophthalmology*. 2008;115(3):546-552.
2. Shields CL, Furuta M, Mashayekhi A, et al. Visual acuity in 3422 consecutive eyes with choroidal nevus. *Arch Ophthalmol*. 2007;125(11):1501-1507.
3. Sumich P, Mitchell P, Wang JJ. Choroidal nevi in a white population: the Blue Mountains Eye Study. *Arch Ophthalmol*. 1998;116(5):645-650.
4. Shields CL, Mashayekhi A, Materin MA, et al. Optical coherence tomography of choroidal nevus in 120 patients. *Retina*. 2005;25(3):243-252.
5. Shields CL, Furuta M, Berman EL. Choroidal nevus transformation into melanoma: analysis of 2514 cases. *Arch Ophthalmol*. 2009;127(8):981-987.
6. Zayit-Soudry S, Moroz I, Loewenstein A. Retinal pigment epithelial detachment. *Surv Ophthalmol*. 2007;52(3):227-243.
7. Lavinsky D, Belfort RN, Navajas E, Torres V, Martins MC, Belfort R Jr. Fundus autofluorescence of choroidal nevus and melanoma. *Br J Ophthalmol*. 2007;91(10):1299-302.
8. Shields CL, Pirondini C, Bianciotto C, Materin MA, Harmon SA, Shields JA. Autofluorescence of choroidal nevus in 64 cases. *Retina*. 2008;28(8):1035-1043.
9. Bird AC. Bruch's membrane change with age. *Br J Ophthalmol*. 1992;76(3):166-168.
10. Elan MJ, Fine SL, Murphy RP, Patz A, Auer C. The natural history of serous retinal pigment epithelum detachment in patients with age-related macular degeneration. *Ophthalmology*. 1986;93(2):224-230.
11. Levy J, Shneck M, Klemperer I, Lifshitz T. Treatment of subfoveal choroidal neovascularization secondary to choroidal nevus using photodynamic therapy. *Ophthalmic Surg Lasers Imaging*. 2005;36(4):343-5.
12. Zografos L, Mantel I, Schalenbourg A. Subretinal choroidal neovascularization associated with choroidal nevus. *Eur J Ophthalmol*. 2004;14(2):123-31.
13. Parodi MB. Transpupillary thermotherapy for subfoveal choroidal neovascularization associated with choroidal nevus. *Am J Ophthalmol*. 2004;138(6):1074-5.
14. Ladas ID, Kotsolis AI, Papakostas TD, Rouvas AA, Karagiannis DA, Vergados I. Intravitreal bevacizumab combined with photodynamic therapy for the treatment of occult choroidal neovascularization associated with serous pigment epithelium detachment in age-related macular degeneration. *Retina*. 2007;27(7):891-6.