Halo Nevus of the Choroid: An Innocent Bystander

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The choroidal nevus classically appears as a brown or slate gray pigmented mass located in the choroid and is without retinal component.[1] This lesion is generally less than 2 mm in thickness.[1] Previous clinical studies have found that the presence of choroidal nevi vary from an estimated 0.2% to 30% of patients.[2] In an analysis of the clinical features of 3422 eyes with choroidal nevi, it was found that, when separately evaluating the young patients (<20 years), mid-adult patients (21-50 years), and older adult patients (>50 years), race was predominantly white in 95%, 98%, and 99%, and sex was female in 63%, 65%, and 63% of patients, respectively.[2] The tumor was located in the extrafoveolar location in 86%, 93%, and 94% of patients and more often in the nasal or temporal quadrant than inferior or superior quadrant.[2] The nevus was pigmented in 89%, 74%, and 77% of patients. From this oncology clinic-based survey, many patients were referred with highly suspicious lesions, and growth into melanoma occurred in 14%, 9%, and 6% of patients, respectively.[2]

There is an interesting variant of choroidal nevus that manifests a central brown color with a yellow surrounding halo, termed "halo nevus".[3] Halo nevi can occur in the skin and in the eye.[4] From a dermatology perspective, a review of 35 patients with cutaneous halo nevi revealed mean age at presentation of 18 years, sex 54% female, and race nearly all white From an ophthalmology perspective, halo nevus represents 5% of all choroidal nevi.[2] Herein, we report a case and further explore this nevus variant.

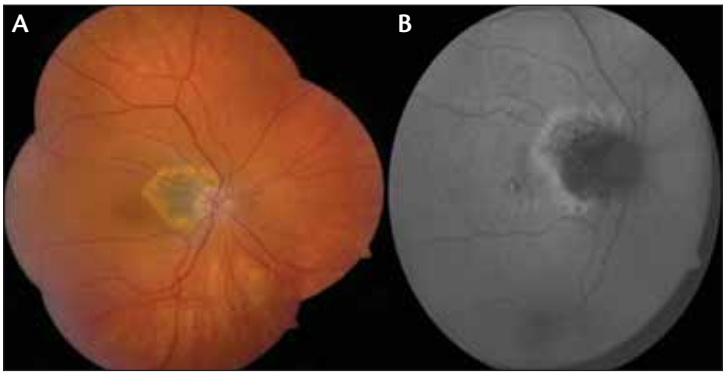

Figure 1. A halo choroidal nevus touching the optic disc showing central brown color and surrounding yellow halo (A). Autofluorescence of halo nevus shows mild hypoautofluorescence of the pigmented portion and slight hyperautofluorescence of the nonpigmented portion (B).

CASE

A 53-year old white woman noted decreased vision in the right eye (OD) for 3 years and a suspicious pigmented choroidal lesion was found. The patient reported no cutaneous or ocular melanoma or autoimmune disease.

On examination, visual acuity was 20/60 OD and 20/20 OS. Intraocular pressures were normal. Bilateral iris freckles were detected. Fundus examination OS was unremarkable. Fundus examination OD disclosed a central brown choroidal mass with a circumferential yellow ring surrounding it. The lesion displayed slightly irregular margins and measured 3.5 x 3.5 x 1.4 mm. There was no overlying subretinal fluid or orange pigment (Figure 1A). Autofluorescence showed mild hypoautofluorescence of the pigmented portion and moderate hyperautofluorescence of the non-pigmented portion. (Figure 1B) Ultrasound revealed a plateau-shaped lesion with thickness of 1.4 mm. These features were consistent with halo choroidal nevus. At 2-year follow-up, the nevus remained stable, with no change in size or color.

COMMENT

Cutaneous halo nevus can appear on the face, scalp, thigh, arm, neck, forearm, and leg, in decreasing frequen-

cy.[5] Most cutaneous halo nevi appear as a solitary finding, but in one analysis 15% of patients had 2 halo nevi and 6% had 3 halo nevi.[5] It has been reported that benign cutaneous halo nevus can evolve to malignant melanoma, and therefore monitoring is warranted.[5]

There have been few publications on choroidal halo nevus. In a large analysis of 3422 cases of choroidal nevi, halo nevus represented 5%.[3] Choroidal halo nevus was found in 2% of young patients (0-20 years), 8% of mid-adults (20-50 years), and 4% of older adults (>50 years).[2] In an analysis of risk factors predictive of growth of choroidal nevus into melanoma, one study found that the presence of a halo around a choroidal nevus was a protective factor that statistically lessened the risk for transformation.[6] In that analysis, the absence of a halo around a nevus promoted a 7 times greater risk for transformation into melanoma compared to a nevus with a halo.[6]

In an analysis of 150 cases of choroidal halo nevi, symptoms such as decreased vision and flashes/floaters were present in 17%.[3] This nevus variant was recognized at median age of 55 years with median size of 6 mm base and 1.6 mm thickness.[3] In this analysis. only 2 patients (1%) showed multifocal halo nevi. Over a mean of 6 years follow up, transformation into melanoma was found in 4% of cases, at a mean interval of 44 months, and often with predictable risk factors.[3,5]

Some investigators speculate that halo nevi of the skin and eye might be related to autoimmune disease or vitiligo.[7] Some publications conjecture a relationship with cutaneous melanoma and favor the halo to be an immune ring of cytotoxic lymphocytes directed to melanocytic cells.[7] There may be a relationship of choroidal halo nevus with pre-existent cutaneous melanoma.[2]

Although the risk for transformation into melanoma is small, it is advised that patients with choroidal halo nevi be monitored biannually and then annually. It is also recommended that all patients be checked by a dermatologist for coincident or undiscovered cutaneous melanoma. Perhaps the halo nevus is an innocent bystander or marker of underlying cutaneous melanoma. ■

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1. Shields, JA, Shields CL. *Intraocular Tumors: An Atlas and Textbook*, 2nd ed. Philadelphia: Lippincott Williams & Wilkins, 2008.
2. Shields CL, Furuta M, Mashayekhi A, et al. Clinical spectrum of choroidal nevi based on age at presentation in 3422 consecutive eyes. *Ophthalmology.* 2008;115:546-52.
3. Shields CL, Maktabi AM, Jahnle E, Mashayekhi A, Lally SE, Shields JA. Halo nevus of the choroid in 150 cases: the 2010 Henry van Dyke Lecture. *Arch Ophthalmol.* 2010;128(7):859-864.
4. Elder D, Elenitsas R. Halo nevus. In Elder D, Elenitsas R, Jaworksy C, Johnson B, eds, *Lever's Histopathology of the Skin*, 8th ed. Philadelphia: Lippincott-Raven: 1997:652-654.
5. Kopf A, Morrill S, Silberberg I. Broad spectrum of leukoderma acquisitum centrifugum. *Arch Dermatology.* 1965;92(1): 14-33.
6. Shields CL Shields JA, Kiratli H, et al. Risk factors for growth and metastasis of small choroidal melanocytic lesions. *Ophthalmology.* 1995;102:1351-1361.
7. Fournier G, Albert D, Wagoner MD. Choroidal halo nevus occurring in a patient with vitiligo. *Surv Ophthalmol.* 1984;28:671-672.
8. Epstein WL, Sagebeil R, Spitler L, Wybran J, Reed W, Blois S. Halo nevi and melanoma. *JAMA.* 1973;225(4):373-377.