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Multifocal Unilateral Orange Fundus Tumors in a Young Man

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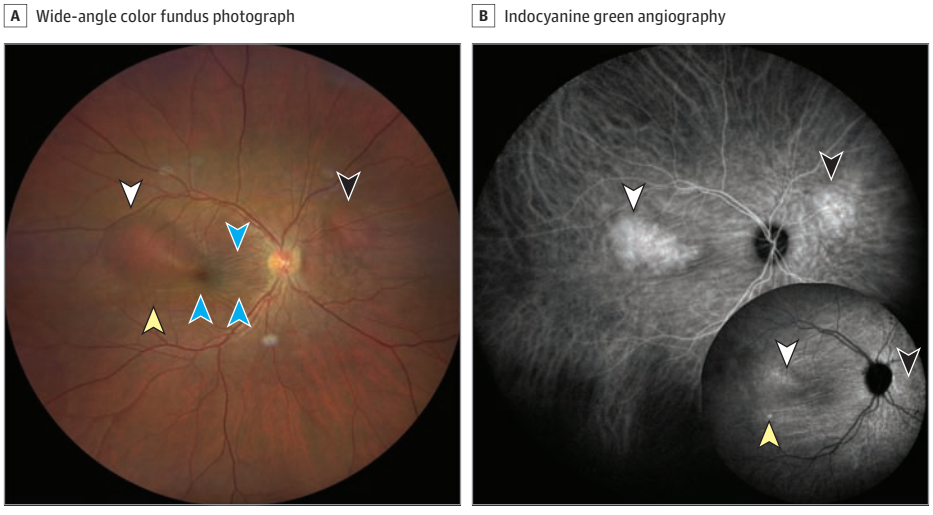

Figure 1. A, Wide-angle color fundus photograph of the right eye showed 2 orange choroidal tumors, with one in the temporal macular region (white arrowhead) and the second in the superonasal peripapillary region (black arrowhead). There was a tiny retinal pigment epithelial detachment (yellow arrowhead) and horizontal macular choroidal folds (blue arrowheads). B, Indocyanine green angiography of the right eye at 181 seconds demonstrated early hyperfluorescence of both tumors (white and black arrowheads). Inset, Late washout of the choroidal tumors (white and black arrowheads) with pooling of dye in the retinal pigment epithelial detachment (yellow arrowhead).

A 38-year-old White man was referred for evaluation of an incidentally discovered asymptomatic macular choroidal mass in the right eye. The spherical equivalent of refractive error was +2.50 D OU. His medical history was noncontributory. Visual acuity was 20/20 OU. External and anterior segment examination findings were unremarkable in each eye. Fundus examination of the right eye revealed an orange mass in the temporal macular region, measuring 6.0 mm × 6.0 mm in basal dimension and 2.6 mm in thickness, abutting the foveola, with prominent horizontal macular choroidal folds and a solitary retinal pigment epithelial (RPE) detachment inferiorly (**Figure 1**A). In addition, another orange mass was identified in the right eye nasal to the optic disc, measuring 3.0 mm × 3.0 mm in basal dimension and 2.1 mm in thickness. Indocyanine green angiography (ICGA) showed early choroidal hyperfluorescence of both tumors, with rapid choroidal washout and pooling of dye within the RPE detachment (Figure 1B). On optical coherence tomography (OCT), both lesions were located within the choroid, with smooth apical surface configuration, choroidal vascular expansion, no subretinal fluid, and nearby choroidal folds. On ultrasonography, both lesions were acoustically dense. Subfoveal choroidal thickness measured 310 µm OD and 252 µm OS. Examination and multimodal imaging of the left eye showed subtle horizontal macular choroidal folds and no mass. Axial length by A-scan ultrasonography was 21.7 mm OD and 22.7 mm OS, consistent with axial hyperopia greater in the right eye than in the left eye.

WHAT WOULD YOU DO NEXT?

A. Transpupillary thermotherapy

B. Laser photocoagulation

C. Imaging of the chest and abdomen

D. Plaque radiotherapy

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Diagnosis

Multifocal circumscribed choroidal hemangioma

What to Do Next

C. Imaging of the chest and abdomen

Discussion

In a series of 5586 amelanotic choroidal tumors in 4638 eyes, the 5 most common diagnoses were choroidal melanoma (41%), choroidal nevus (21%), choroidal metastasis (16%), choroidal hemangioma (8%), and peripheral exudative hemorrhagic chorioretinopathy (4%).[1] In that analysis, tumor diagnosis was dependent on patient age, sex, and tumor basal diameter, and a diagnostic flowchart for amelanotic choroidal tumors was constructed based on these features.[1] Given the features in this case, including the patient being aged 38 years, being male, and having a basal dimension less than 8 mm, the 3 most common diagnoses were choroidal hemangioma (34%), nevus (20%), or melanoma (19%).[1] Features against choroi-

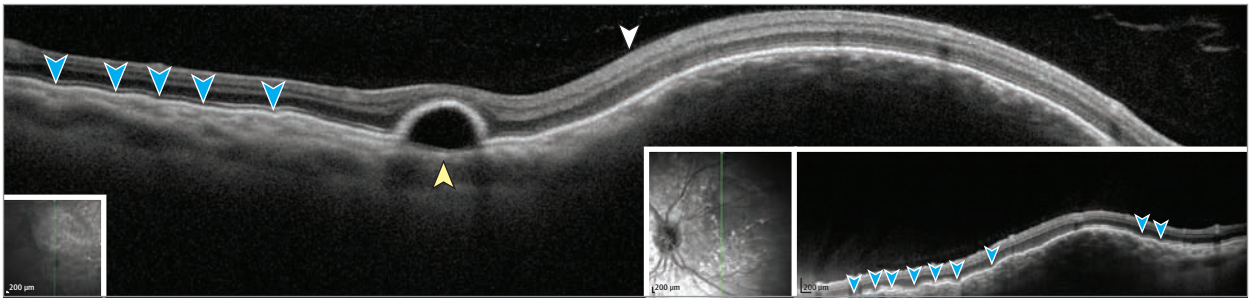

Figure 2. Optical coherence tomography of the right eye with a vertical scan through the macula showing choroidal folds (blue arrowheads), retinal pigment epithelial detachment (yellow arrowhead), and choroidal hemangioma (white arrowhead) with intact choroidal vascular detail and no subretinal fluid. Inset, Scan through the superonasal, apically smooth, dome-shaped lesion shows adjacent choroidal folds (blue arrowheads).

dal melanoma in this case included orange tumor color, multifocality, and ultrasonographic acoustic solidity. Features against choroidal nevus included orange tumor color and lack of overlying RPE alterations. Neither melanoma nor nevus would likely show early hyperfluorescence on ICGA. Features supporting the diagnosis of choroidal hemangioma included the orange tumor color, ultrasonographic acoustic solidity, and hyperfluorescence on ICGA with late washout (Figure 1). Furthermore, OCT features of a smooth, dome-shaped choroidal mass with expansion of choroidal vascular markings (**Figure 2**) are suggestive of choroidal hemangioma.[2] The only atypical finding in this case was the presence of 2 choroidal hemangiomas in 1 eye.

Circumscribed choroidal hemangioma typically presents as a unilateral, solitary, orange mass. Given the multifocality in this case, choroidal metastases were considered, but with the need to rule out orange choroidal metastases, such as lung carcinoid tumor, renal cell carcinoma, and thyroid carcinoma.[3,4] Thus, systemic evaluation and imaging of the chest and abdomen was the correct choice (option C). Computed tomography showed no primary tumors elsewhere.

Multifocal circumscribed choroidal hemangioma is rare, with only 2 reported cases in the literature.[5,6] This benign vascular mass is a presumed-congenital malformation and is typically diagnosed at a mean age of 52 years, especially when symptomatic.[7] Based on 2 large cohort studies of 200 cases and 458 cases, multifocality of circumscribed choroidal hemangioma was found in only 1 case (0.2%) and bilaterality in none (0%).[7,8]

Sobol et al[9] found increased subfoveal choroidal thickness in the contralateral eye of patients with circumscribed choroidal hemangioma vs age- and gender-matched controls. This was suggestive of a more diffuse process and possibly bilateral alterations in choroidal vasculature in affected patients. Some speculate choroidal hemangioma is on the spectrum of pachychoroid disease,[9] which could explain the RPE detachment, a feature of pachychoroidopathy.[10]

Treatment for circumscribed choroidal hemangioma is provided if there is related subfoveal fluid or intraretinal foveal edema affecting visual acuity. The treatment of choice is photodynamic therapy.[7] Other options that were not necessary in this asymptomatic case included transpupillary thermotherapy (option A), laser photocoagulation (option B), or plaque radiotherapy (option D).

Patient Outcome

The patient was monitored annually for 6 years. Visual acuity was stable, the fovea remained intact on OCT, and there was no change in either hemangioma.

ARTICLE INFORMATION

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