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Orange pigment overlying a choroidal tumor

Case

A 55-year-old Hispanic lady presented for an opinion regarding decreased visual acuity in her right eye (OD) for 6 months. She had no relevant background and was of Spanish descent. A previous ophthalmic consult had resulted in a presumptive diagnosis of choroidal melanoma and enucleation was suggested. On our examination, best-corrected visual acuity (BCVA) was 20/100 OD and 20/20 in her left eye (OS). Anterior segment examination of OD was normal; however, the fundus examination revealed a 5 \times 5 mm subretinal lightly pigmented lesion under the fovea and extending inferiorly. Overlying orange pigment and subretinal fluid were clinically visualized, and later confirmed by fundus photography [Fig. 1a] and fundus autofluorescence (FAF) [Fig. 1b]. The OS examination was unremarkable. Ancillary studies were ordered [Fig. 1c and d].

What is your next step?

- A) Enucleation and histopathological analysis
- B) Diagnostic incisional biopsy via transvitreal approach
- C) Plaque brachytherapy
- D) Photodynamic therapy (PDT) with verteporfin.

Findings

Fundus photography OD showed the presence of orange pigment over the lesion [Fig. 1a, white arrow] with moderate overlying hyperautofluorescence and marked surrounding hyperautofluorescence on FAF [Fig. 1b]. Optical coherence tomography (OCT) evidenced a dome-shaped mass with intralesional expanded vascular pattern within the choroid and overlying subretinal fluid [Fig. 1c, black arrow]. Ocular ultrasound (US) showed an echodense, dome-shaped choroidal mass measuring 2.67 mm in thickness [Fig. 1d]. Additional workup with indocyanine green (ICG) angiography showed early hypercyanescence of the mass in the pre-arterial phase and diffuse late staining of the lesion. A diagnosis of circumscribed choroidal hemangioma (CCH) was confirmed and PDT was advised.

Diagnosis:

Circumscribed Choroidal Hemangioma

Correct answer: D. Photodynamic therapy (PDT) with verteporfin

Discussion

Clinical findings and multimodal imaging strongly suggested the diagnosis of benign CCH; hence, enucleation (answer A) was not necessary. Intraocular biopsy (answer B) could produce intraocular hemorrhage and is not advisable with this tumor. Plaque radiotherapy (answer C) can be an effective treatment for CCH, but PDT is currently the treatment of choice with a safer side-effect profile. [11]

Choroidal hemangioma is a benign vascular tumor of the uveal tract and can be circumscribed or diffuse. [1-2] More than 90% of reported cases are found in patients in the fifth decade of life, with blurred vision being the most common symptom. [1-2] The clinical characteristics of CCHs include subretinal fluid (72-81%), retinal pigment epithelial hyperplasia (33%) or metaplasia (20%), and macular edema (17-24%). [1-2] Orange pigment can occur over this tumor, but often is not clinically visible as it blends into the orange color of the mass. A large series on pseudomelanomas revealed that CCH is the fifth most common lesion misdiagnosed as choroidal melanoma. [3]

Treatment options for CCHs include observation (if asymptomatic and without subretinal fluid), photodynamic therapy (PDT), plaque brachytherapy, and lens-sparing external beam radiotherapy (LS-EBRT). ^[1] PDT is strongly preferred over radiation methods as this avoids long-term radiation complications. An analysis of 458 patients with choroidal hemangioma in the pre-PDT era (vs. PDT era) revealed the final visual outcome at 20/400 (pre-PDT) vs 20/63 (PDT) over a 51-year follow-up. ^[2]

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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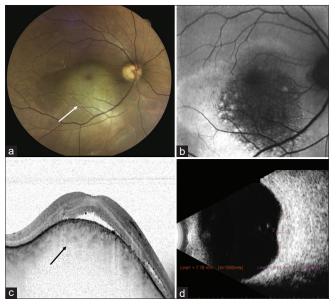


Figure 1: Fundus photography OD showed the presence of orange pigment over the lesion (a, white arrow) with moderate overlying hyperautofluorescence and marked surrounding hyperautofluorescence on FAF (b). Optical coherence tomography (OCT) evidenced a dome-shaped mass with intralesional expanded vascular pattern within the choroid and overlying subretinal fluid (Figure 1c, black arrow). Ocular ultrasound (US) showed an echodense, dome-shaped choroidal mass measuring 2.67 mm in thickness (d)

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Conflicts of interest

There are no conflicts of interest.

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