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Clinical challenges

This, that, or something different?

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(In keeping with the format of a clinical pathological conference, the abstract and key words appear at the end of the article.)

1. Case report

A 72-year-old man was referred for the evaluation of a single choroidal mass in his right eye that was discovered incidentally on dilated examination. On presentation, the patient had no visual complaints and his visual acuity was 20/20 in both eyes. His pupillary examination, intraocular pressures, and anterior segment examination results were within normal limits in both eyes. Posterior segment examination of the left eye was normal. Posterior segment examination of the right eye revealed an elevated dome-shaped yellow-brown choroidal mass measuring about 10 × 10 mm and located in the inferonasal quadrant (Fig. 1). The mass was associated with overlying retinal pigment epithelium changes. There was no evidence of drusen, orange pigment, hemorrhage, or subretinal fluid.

What is your initial clinical impression?

2. Comments

2.1. Comments by Carol Shields, MD

On the basis of wide-angle fundus imaging, this patient demonstrated a slightly pigmented, abruptly elevated choroidal mass inferior to the macular region, measuring approximately 10 mm in basal dimension and estimated 4 mm in thickness. There was no evident subretinal fluid. There were retinal pigment epithelial alterations overlying the mass, but these alterations were visible elsewhere in the fundus, so they might not necessarily be related to the mass. There was no sign of choroidal melanocytosis. These features are most suggestive of choroidal melanoma, but one must rule out simulators of melanoma including nevus, peripheral hemorrhagic chorioretinopathy, metastatic tumor, and others.[7,8]

3. Case report (continued)

Further questioning revealed that the patient's previous dilated fundus examination was approximately 5 years ago with no note of an intraocular mass. He denied any recent illnesses, travel, or exposure.

He has a history of 2 primary tumors: multiple myeloma and neuroendocrine thymoma diagnosed 6 and 10 years before presentation, respectively. The neuroendocrine thymoma has metastasized to the lung and pancreas requiring

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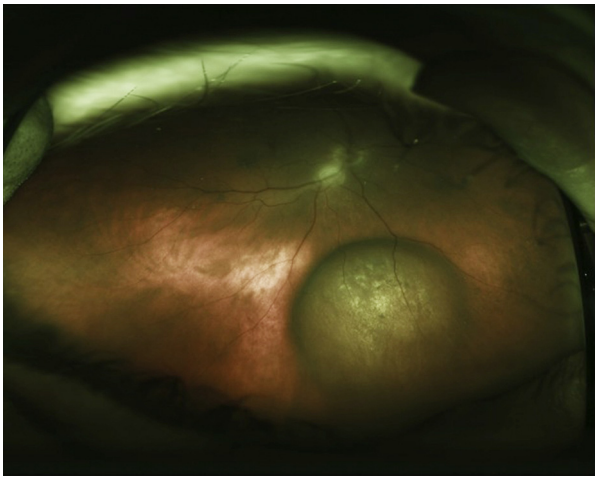

Fig. 1 – Wide-field colored fundus photograph of the right eye revealing a dome-shaped inferonasal yellow or orange tumor with overlying retinal pigment epithelium changes.

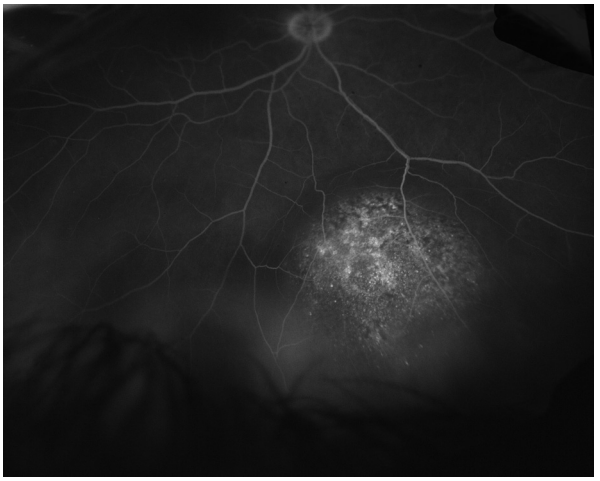

Fig. 2 – Late-frame (7 minutes 22 seconds) wide-field fluorescein angiography of the right eye showing stippled hyperfluorescence without associated overlying vascular leakage or presence of intrinsic vessels.

partial pancreatectomy 2 years ago. The lung metastasis is stable and being monitored without treatment for the past 3 years. The multiple myeloma is in remission. The patient is currently taking lenalidomide, dexamethasone, and enoxaparin.

What would you order next?

4. Comments (continued)

4.1. *Comments by Dr. Shields*

The history of two previous malignancies adds an interesting twist to this case and raises suspicion for possible choroidal metastasis from either the neuroendocrine thymoma or myeloma. Choroidal infiltration by myeloma is termed "plasmacytoma." Intrinsic pigment within a choroidal mass is generally suggestive of a melanocytic tumor such as nevus or melanoma, but occasionally, metastatic neuroendocrine tumors can have a slightly pigmented hue. Plasmacytoma is usually sessile and ill defined, unlike this tumor, and with an orange-yellow hue. At this point, diagnostic testing with ultrasonography, optical coherence tomography, fundus autofluorescence, and fluorescein angiography might be helpful. However, given the medical history, fine-needle aspiration biopsy (FNAB) might ultimately be necessary.

What is your working differential diagnosis?

5. Comments (continued)

5.1. *Comments by Dr. Shields*

Fluorescein angiography (Fig. 2) revealed a minimally fluorescent choroidal mass with overlying "window defects" from retinal pigment epithelial alterations. There was no "double circulation" to suggest choroidal melanoma. Ocular ultrasonography (A-scan; Fig. 3) showed moderate to high internal reflectivity with slight angle kappa, suggestive of metastasis or melanoma. B-scan ultrasonography (Fig. 4) disclosed a slightly hollow choroidal mass of 4 mm thickness and without overlying subretinal fluid, consistent with either melanoma or metastasis. At this point, the imaging features could be suggestive of either choroidal melanoma or metastasis, and other considerations like choroidal hemangioma or inflammatory conditions would be unlikely.

6. Case report (continued)

A discussion with the patient's hematologist and oncologist revealed that his metastatic neuroendocrine thymoma had been stable for the past 3 years. Although his multiple myeloma was now in remission, there had been episodes of biopsy-proven extramedullary plasmacytoma. They believed that both primary tumors could be the cause of the choroidal metastasis. They also stated that both primary tumors were indolent, and the patient had a favorable overall prognosis for survival.

What would you do next?

7. Comments (continued)

7.1. *Comments by Dr. Shields*

On the basis of this information, with two primary malignancies and possible choroidal metastasis, we would advise FNAB. We would approach this tumor via a trans pars plana, transvitreal route using a 27-gauge needle on a connector tubing and attached to a 10 mL syringe. After aspiration, the cells should be stored in a cytologic preservative to protect cellular integrity and avoid cytolysis. Prompt evaluation of the specimen by a qualified cytopathologist is important for accurate diagnosis.