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Case report

A rare presentation of choroidal metastasis from primary esophageal adenocarcinoma successfully treated with intensity-modulated radiation therapy

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ABSTRACT

Purpose: In this case report, we present a case of choroidal metastasis from a primary esophageal adenocarcinoma that was treated successfully with intensity-modulated radiation therapy.
Observations: A 65-year-old male with known stage IV esophageal adenocarcinoma presented with a central scotoma in his left eye and was ultimately found to have a large choroidal metastatic lesion with overlying subretinal fluid. IMRT was administered over the course of four weeks, resulting in restoration of the patient's vision, regression of the metastatic lesion, and resolution of the subretinal fluid. As of 16 months following completion of radiation, there remains no evidence of choroidal recurrence or radiation-associated ocular complications.
Conclusions: and Importance: To our knowledge, this is the first published case report of a choroidal metastasis from esophageal cancer responding durably to IMRT. IMRT should therefore be considered a viable treatment option for this rare disease.

1. Introduction

The choroid is an uncommon but well-documented site for metastasis of solid tumors, most commonly breast and lung carcinomas. On autopsy, choroidal metastasis can be found histologically in 0–11% of patients with any known metastatic malignancy and 4–12% of patients with breast and lung cancer.[1–4] However, choroidal metastases originating from the esophagus are exceedingly rare. In fact, a recent review of multiple case series found that only 8 of 918 cases (0.87%) of choroidal metastases were attributed to an esophageal origin.[5] In this report, we summarize the presentation and management of a patient who developed a choroidal metastasis from primary esophageal adenocarcinoma and then review the limited published literature pertaining to this rare clinical entity.

1.1. Case report

A 65-year-old Caucasian male was diagnosed with Her-2 positive, locally advanced adenocarcinoma of the lower esophagus and treated definitively with chemotherapy, radiation, and Ivor-Lewis esophagectomy. Eighteen months later, his cancer recurred in bone, non-regional lymph nodes, and the surgical bed. Palliative radiation to an iliac metastasis and systemic chemoimmunotherapy (capecitabine, oxaliplatin, trastuzumab) were initiated, and subsequent imaging showed response to therapy.

Ten months into treatment, however, the patient noticed a central scotoma in his left eye and was referred to an ophthalmologist who reported visual acuity of 20/20 in the right eye and 20/25 in the left eye without an afferent pupillary defect. Dilated funduscopic examination revealed a large, elevated yellow choroidal lesion in the temporal macula with RPE changes and overlying subretinal fluid (Fig. 1a). Spectral-domain optical coherence tomography (SD-OCT) revealed subretinal fluid overlying the area of choroidal infiltration with shaggy photoreceptors and RPE debris, as well as irregular foveal contour caused by the choroidal mass (Fig. 1b and c). Fluorescein angiography demonstrated window defects in areas of RPE atrophy

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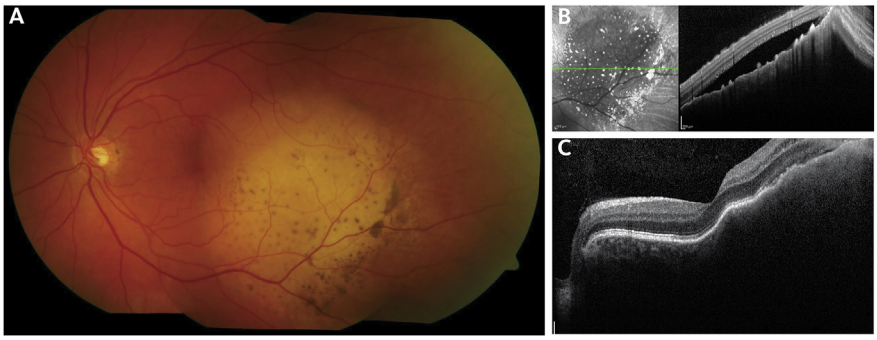

Fig. 1. (a) Color fundus photograph of the left eye revealing a yellow, creamy, elevated lesion in the posterior pole with overlying RPE changes; (b) Spectral-domain optical coherence tomography (SD-OCT) of the left eye through the lesion demonstrating choroidal elevation and infiltration with overlying subretinal fluid; (c) SD-OCT through the fovea demonstrating involvement of the choroid centrally causing irregular foveal contour. (For interpretation of the references to colour in this figure legend, the reader is referred to the web version of this article.)

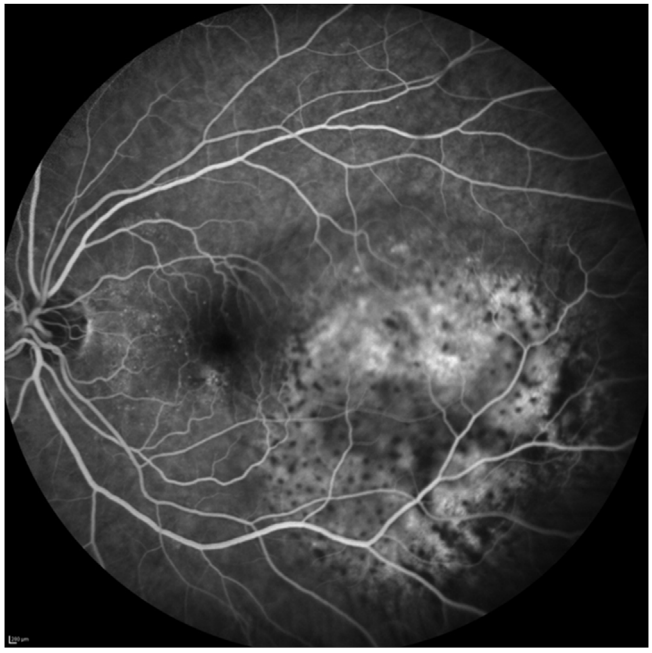

Fig. 2. Fluorescein angiography of the left eye revealing window defects in areas of RPE atrophy interspersed with blockage from RPE clumping.

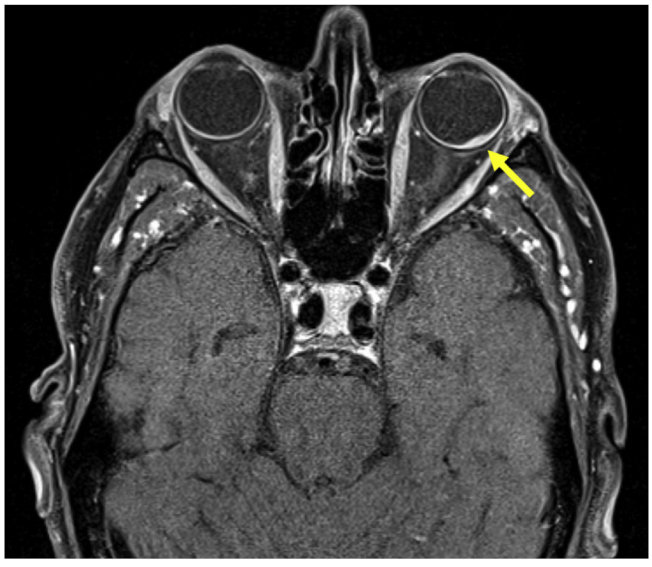

Fig. 3. Pre-treatment T1-weighted MRI of the orbits, showing a 6 mm × 2 mm elevated enhancing lesion on the left posterolateral globe (arrow).

interspersed with blockage from RPE clumping (Fig. 2). Standardized A- and B-scan ultrasound demonstrated a choroidal mass with a largest basal dimension of 11.4 mm and an apical height of 3.4 mm with medium-high internal reflectivity. These findings were consistent with the clinical suspicion of choroidal metastasis. Neuroimaging including MRI (Fig. 3) revealed a flat lesion in the posterolateral left globe, but no evidence of intracranial lesions. The patient's multidisciplinary team recommended that he undergo targeted ocular therapy while continuing systemic chemotherapy.

The patient received 40 Gy of intensity-modulated radiation therapy (IMRT) in 20 fractions over a course of four weeks (Fig. 4) concurrently with radiosensitizing 5-fluorouracil and trastuzumab. The patient responded well, with resolution of the subretinal fluid, regression of the metastatic lesion (Fig. 5), and restoration of visual acuity (20/20) after the completion of IMRT. After 16 months of follow-up, there is no evidence of clinical or radiographical choroidal recurrence or radiation-induced toxicity. Repeat neuroimaging one year after treatment showed resolution of the previously noted choroidal mass (Fig. 6).

2. Discussion

To our knowledge, this is the first published report of a choroidal metastasis from primary esophageal cancer that responded in a complete and durable manner to radiotherapy. While esophageal cancer metastasizing to the choroid is exceedingly rare, choroidal metastases are the most common intraocular malignancy.[5] A substantial portion of patients with choroidal metastases may be asymptomatic, but common presenting symptoms include blurred vision, metamorphopsia, floaters, and photopsias. Visual acuity at presentation can vary, depending on the extent of the lesions and the presence or absence of subretinal fluid. When Shields et al.[6] examined 520 eyes in 420 patients with metastatic uveal lesions, they localized 88% of metastatic lesions to the choroid, while iris and ciliary body lesions were far less common.[6] The propensity of tumor cells to seed the choroid preferentially likely stems from its rich vascular supply.

Uveal metastases are most commonly attributed to primary tumors of the breast and lung.[2,7–9] Only 4% of uveal metastases arise from the gastrointestinal tract, and only a small percentage of these originate in the esophagus.[6,10–19] Furthermore, most reported cases of esophageal cancer metastasizing to the uvea are of squamous histology. Only a few cases of uveal metastases from adenocarcinoma of the esophagus have been reported in the literature.[10,11,15–18]

Management of choroidal metastases depends on a number of factors, including primary tumor site, tumor histology, extent and burden of extra-uveal disease, the patient's underlying health, overall prognosis, and symptomatology. Once choroidal metastases have arisen, prognosis is generally poor, so the goal of treatment is salvaging vision in a minimally-invasive manner.[5] Whether or not systemic chemotherapy alone can effectively treat choroidal metastases is debatable, but most agree that local therapy is generally warranted.[5,20] Standard fractionated external beam radiotherapy (EBRT) is the modality most commonly used to treat choroidal metastases, yielding tumor regression