

Metastatic Esophageal Cancer Presenting as an Orbital Mass

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ABSTRACT

We report a case of adenocarcinoma of the esophagus presenting as an orbital metastasis prior to the primary diagnosis. A 66-year-old white male presented to his ophthalmologist with right orbital swelling for several months. Magnetic resonance imaging revealed a supraorbital infiltrative mass. Pathology from the mass revealed findings consistent with adenocarcinoma of gastrointestinal origin. Upper endoscopy revealed distal esophageal stricture and irregularities. Pathology from the esophagus showed the same malignancy found in the orbit. An orbital swelling can manifest as the initial presentation of metastatic disease and should be taken seriously to avoid delay in diagnosis and treatment.

INTRODUCTION

Malignant orbital masses occur rarely. The 3 most common primary sites to metastasize to the orbit are prostate, breast, and lung.^{1,2} A recent large retrospective series identified that the rate of orbital metastasis was 3%.¹ Orbital masses are usually slow growing and are often followed up as an outpatient; in certain circumstances, however, they can manifest as the first sign of systemic illness. In as many as 19% of the cases, these are the only manifestation of underlying malignancies and can easily be overlooked.^{3,4}

CASE REPORT

A 66-year-old male was sent to our emergency department by his ophthalmologist for the evaluation of a right orbital mass and proptosis. On follow-up at 3 months, magnetic resonance imaging showed an infiltrative mass on the right supraorbital region with mass effect on the patient's right globe (Figure 1). The patient did not have any visual complaints but had noticed swelling for the last 3 months, which had progressively worsened. He also complained of mild dysphagia with nearly 18-kg weight loss in 3 months. He underwent right orbitotomy and resection of the orbital mass. Immunohistochemical staining performed on the tumor cells was positive for CK7, MUC5AC, and ARG2 and negative for CK5/6, P63, Napsin A, and TTF1, suggesting adenocarcinoma of upper gastrointestinal origin (Figure 2).

Chest, abdominal, and pelvic computed tomography revealed irregular and thickened distal esophageal tissue. The patient then underwent upper endoscopy, which revealed stricture of the distal esophagus, from which multiple biopsies were taken (Figure 3). The biopsy revealed the same malignancy, which confirmed that the mass was poorly differentiated esophageal adenocarcinoma (Figure 4). Chest and abdominal computed tomography and brain magnetic resonance imaging showed no other metastases. This was later confirmed with positron emission tomography, which showed uptake in the right orbit apart from the esophagus. The

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Figure 1. Brain magnetic resonance imaging (T2W) showing a heterogeneous infiltrative mass (arrow) within the right superior orbit measuring approximately 3.3 x 3.9 x 2.0 cm.

patient underwent placement of an esophageal stent for dysphagia and was discharged home with outpatient follow up with oncology. Positron emission tomography was planned as an outpatient.

DISCUSSION

Orbital swelling and proptosis are the most common manifestations of primary eye malignancy.^{3,4} The incidence of an

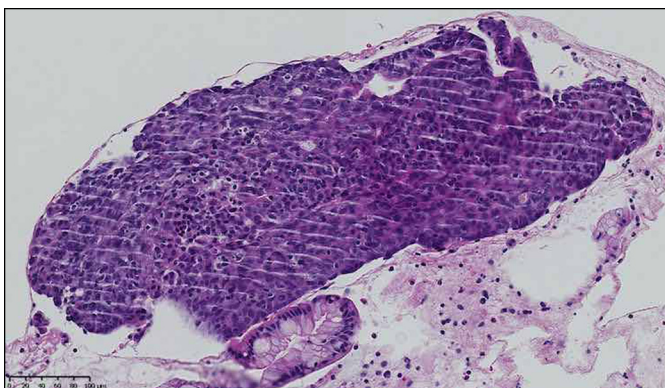


Figure 2. Orbital mass biopsy showing histomorphology consistent with adenocarcinoma of gastrointestinal origin (hematoxylin and eosin staining, 200x).



Figure 3. Upper endoscopy showing distal esophageal stricture.

isolated orbital lesion being a metastatic disease is rare.^{2,5,6} In one of the largest reported series of orbital tumors, metastases accounted for 7% of the total orbital tumors in adults.^{1,2} Of these, the most common primary sites were breast and prostate, accounting for 48% and 12% of the total orbital metastases, respectively.^{1,2} Esophageal and urothelial carcinoma metastases to the orbit have been reported, but they occurred at a much lower frequency.^{5,6} Upon our review of the literature we found very few reported cases of esophageal carcinoma with metastasis to the orbit.^{2,5-7} The typical presentation is with a mass lesion causing proptosis or an ocular motility defect.^{4,6} There have been some reported cases of orbital metastases that present as orbital cellulitis or abscess.¹

Survival after diagnosis of orbital metastases ranges from 13 months to 1.5 years, with variation due to histologic type.⁸⁻¹⁰ For instance, orbital carcinoid metastases appears to have a median survival of 24 months in one series.¹¹ No well-defined

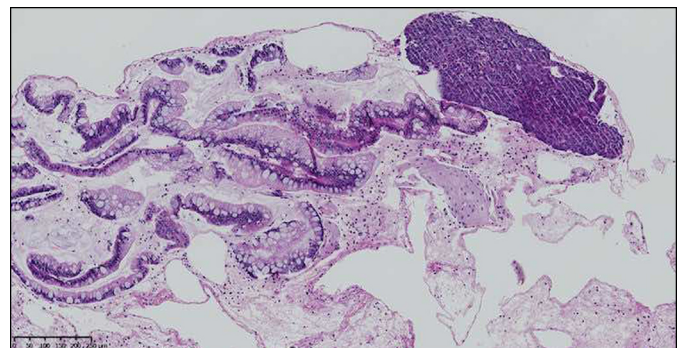


Figure 4. Esophageal biopsy showing a single nest of tumor cells in a background of intestinalized metaplastic epithelium (hematoxylin and eosin staining, 80x).

treatment modality exists for orbital metastasis. Options include radiotherapy, chemotherapy, hormone therapy, surgery, and immunotherapy.¹⁰ For example, neuroendocrine tumors metastatic to the orbit can be treated with stereotactic radiotherapy,¹² radiolabeled octreotide, or with peptide-receptor radiolabeled therapy using ¹⁷⁷Lu-DOTATATE.¹³

DISCLOSURES

Author contributions: S. Pokharel wrote the manuscript and is the article guarantor. SJ Richter reviewed and edited the manuscript. G. Kabbach edited the manuscript and obtained the figures. L. Chiu obtained endoscopy slides.

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Informed consent was obtained for this case report.

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