A Common Procedure Leads to the Diagnosis of an Uncommon Cause of Dysphagia

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INTRODUCTION

Epithelioid angiosarcoma is a rare and aggressive soft-tissue endothelial cell neoplasm that accounts for less than 1% of sarcomas and rarely occurs in the gastrointestinal tract.

CASE PRESENTATION

A 41-year-old female without prior medical history who presented with progressive dysphagia and unintentional weight loss. Initial upper endoscopy found to have a narrowed gastroesophageal junction (GEJ) that required balloon dilation to 10mm and the use of a pediatric upper endoscope to traverse. Only thickened gastric folds at the cardia were seen, which were biopsied, see Figure 1. Histology revealed chronic active moderate gastritis with Helicobacter pylori and treated with triple therapy. However, due to ongoing dysphagia a contrast enhanced computed tomography (CT) of the chest was obtained and demonstrated thickening of the distal esophageal wall. Next, an endoscopic ultrasound (EUS) was performed and demonstrated unusual thickening of the distal esophageal wall, which underwent fine needle biopsy (FNB), see Figure 2. Histologically, there was cellular proliferation of oval and epithelioid cells arranged in sheets and clusters on hematoxylin and eosin (H&E), see Figure 3a. Immunohistochemically, the tumor was positive for ERG, a nuclear stain for vascular differentiation (Figure 3b), positive for CD34, which stains endothelial cells (Figure 3c) and cytokeratin (CK) staining of the mucosa (Figure 3d), another marker for epithelial cells. The overall features with supporting immunoprofile were consistent with high-grade epithelioid angiosarcoma of the distal esophagus. She was treated with neoadjuvant chemo- and radiotherapy followed by a robotic laparoscopic lvor Lewis esophagectomy, see Figure 4. Two months post-operatively, she has been disease free and tolerating a soft diet.



Figure 1. Thickened gastric folds at the cardia on retroflexion.



Figure 3. (a) H&E, (b) ERG, (c) CD34, (d) CK.



Figure 2. EUS and FNB of distal esophageal tumor with irregular boarder extending into adjacent tissue.



Figure 4. Gross photograph of the distal esophagus and proximal stomach resection with tumor at the distal esophagus/gastroesophageal junction

DISCUSSION

The prognosis of angiosarcomas has typically been that of an aggressive cancer with a poor prognosis.

One case report in China described a patient that presented with dysphagia and weight loss that was found to have esophageal epithelioid angiosarcoma. This patient had an intraluminal polypoid lesion with pulmonary metastasis and died one month later.

This case highlights the importance of timely recognition and diagnosis of dysphagia. Crosssectional imaging in addition to EUS/FNB should be performed to evaluate for less common entities. Multidisciplinary involvement was crucial in providing this patient with an accurate diagnosis and treatment.

SOURCES

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