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CASE REPORT | ESOPHAGUS

Management of Esophageal Inflammatory Myofibroblastic Tumor With Endoscopic Submucosal Dissection

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ABSTRACT

Inflammatory myofibroblastic tumors are rare tumors that have been described in virtually all organs. Even though they are extremely rare in the esophagus, several cases have been described in the literature. Surgical resection has been the therapeutic modality used in most of those cases. In this report, we describe a case of inflammatory myofibroblastic tumor that was successfully managed endoscopically for the first time with the endoscopic submucosal dissection technique.

KEYWORDS: inflammatory myofibroblastic tumor; plasma cell granulomas; esophagus; endoscopic submucosal dissection; esophageal tumor

INTRODUCTION

Inflammatory myofibroblastic tumors (IMTs), also known as plasma cell granulomas, are mesenchymal tumors that mostly occur in young adults with slight male predominance.[1] IMT is composed of myofibroblatic spindle cells with inflammatory cell infiltrates.[1] IMTs most commonly occur in the lungs; however, it has been reported in other organs such as the brain, breasts, thyroid gland, heart, trachea, spleen, kidneys, liver, stomach, colon, ampulla of Vater, and omentum.[2] Even though IMT is extremely rare in the esophagus, few cases have been reported. We report a case of IMT that was successfully resected endoscopically with endoscopic submucosal dissection (ESD).

CASE REPORT

A 76-year-old man underwent esophagogastroduodenoscopy (EGD) for evaluation of anemia (hemoglobin of 7.2 g/dL) at a local hospital on December 17, 2020. EGD showed a mass in the distal esophagus. Biopsies showed spindle cell lesion compatible with an IMT. The patient was referred to our institution for further management. On physical examination, the patient appeared pale, but otherwise, the examination was insignificant. EGD with endoscopic ultrasound was performed on February 8, 2021. A 2 cm esophageal tumor was noted in the distal esophagus on the anterior wall (Figure 1). Sonographically, the lesion was hypoechoic measuring approximately 1.7 cm wide and 0.9 cm deep. There was a focal invasion into the muscularis propria (MP) (Figure 1). Anemia was attributed to the tumor as no other source of bleeding was noted on extensive evaluation. In addition, the patient subsequently developed mild but progressive dysphagia for solid food. After discussion with the surgical oncologist, the patient, and the family, decision was made to proceed with ESD as the patient refused surgical esophagectomy. Given the concern of focal MP invasion on endoscopic ultrasound, the risk of incomplete resection and/or perforation was explained to the patient clearly. On March 3, 2021, EGD with ESD was performed using a ERBE hybrid knife (Figure 2). After marking the tumor with cautery, a mixture of normal saline with methylene blue was used to lift the tumor. Then, circumferential incision was made followed by submucosal dissection till the entire tumor was resected. No traction was needed during the procedure. There was focal attachment to circular fibers of the muscularis propria. Focal dissection of superficial muscularis propria was performed, and the entire lesion was resected en bloc. Three hemoclips were placed at the superficial muscle injury areas. Triamcinolone 10 mg/mL was injected to prevent esophageal stricture. Pathology showed IMT with tumor-free margins (Figure 3). The patient was placed on high-dose proton pump inhibitor and sucralfate. Three

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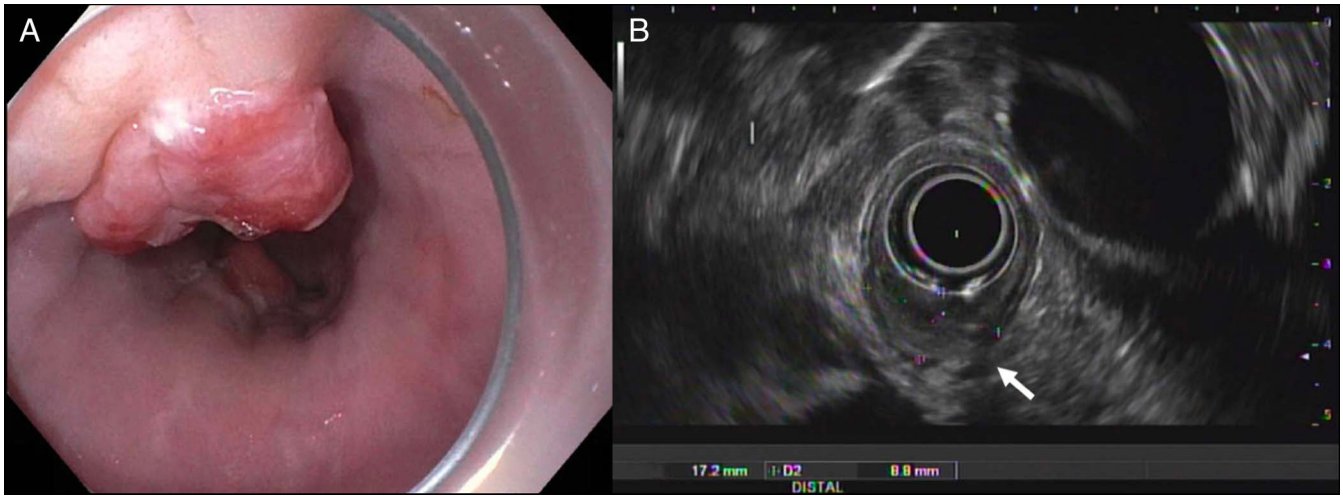

Figure 1. (A) Esophageal tumor noted on EGD that was performed for anemia. (B) EUS evaluation of the esophageal tumor showing partial invasion into the muscularis propria (arrow). EGD, endoscopic submucosal dissection; EUS, endoscopic ultrasound.

months later, the patient presented with dysphagia, and repeat EGD on June 10, 2021, showed mild stricture and small nodule at the ESD site. Dilation up to 18 mm was successful using a controlled radial expansion balloon dilator. Adequate mucosal renting was noted. Biopsies from the nodule showed benign squamous mucosa with granulation tissue. Repeat EGD was performed on March 10, 2022, and showed a subtle stricture in the distal esophagus without evidence of recurrent tumor (Figure 2). The stricture was dilated to 20 mm with adequate mucosal renting. Biopsies were obtained from the ESD site showing benign squamocolumnar mucosa.

DISCUSSION

Although esophageal IMTs are extremely rare, however, several cases have been described in the literature.[2–18] The first case of esophageal IMT was described in 1990.[16] Although those tumors were once believed to be benign, now they are considered intermediate tumor with potential risk of recurrence.[8] Patients with esophageal IMT frequently present with progressive dysphagia (as in our patient), substernal chest pain, weight loss, chronic bleeding with iron deficiency anemia, and/or fever (due to cytokine release).[8] Pathologic evaluation is essential to confirm the diagnosis and will show myofibroblastic spindle cells accompanied by inflammatory infiltration of plasma cells, and lymphocytes (Figure 3). Immunohistochemistry shows positive vimentin, smooth muscle actin, and muscle-specific actin and negative myogenin, myoglobin, CD34, CD117, and S100.[4] More than half of IMTs stain positive for ALK, which is not specific but can be associated with metastasis and recurrence.[4] Surgical resection is considered the standard of care for esophageal IMTs. All cases reported in the literature were managed surgically

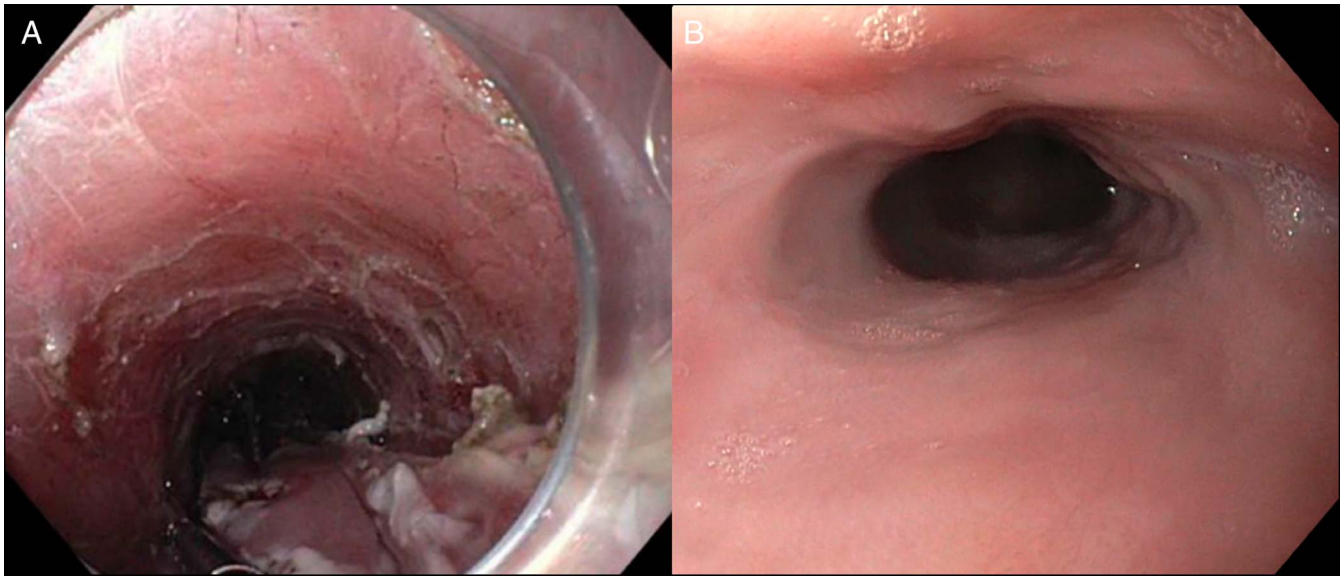

Figure 2. (A) Post-ESD defect with no evidence of residual tumor. (B) One-year follow-up EGD showing a subtle stricture in the distal esophagus without evidence of tumor recurrence. EGD, endoscopic submucosal dissection.

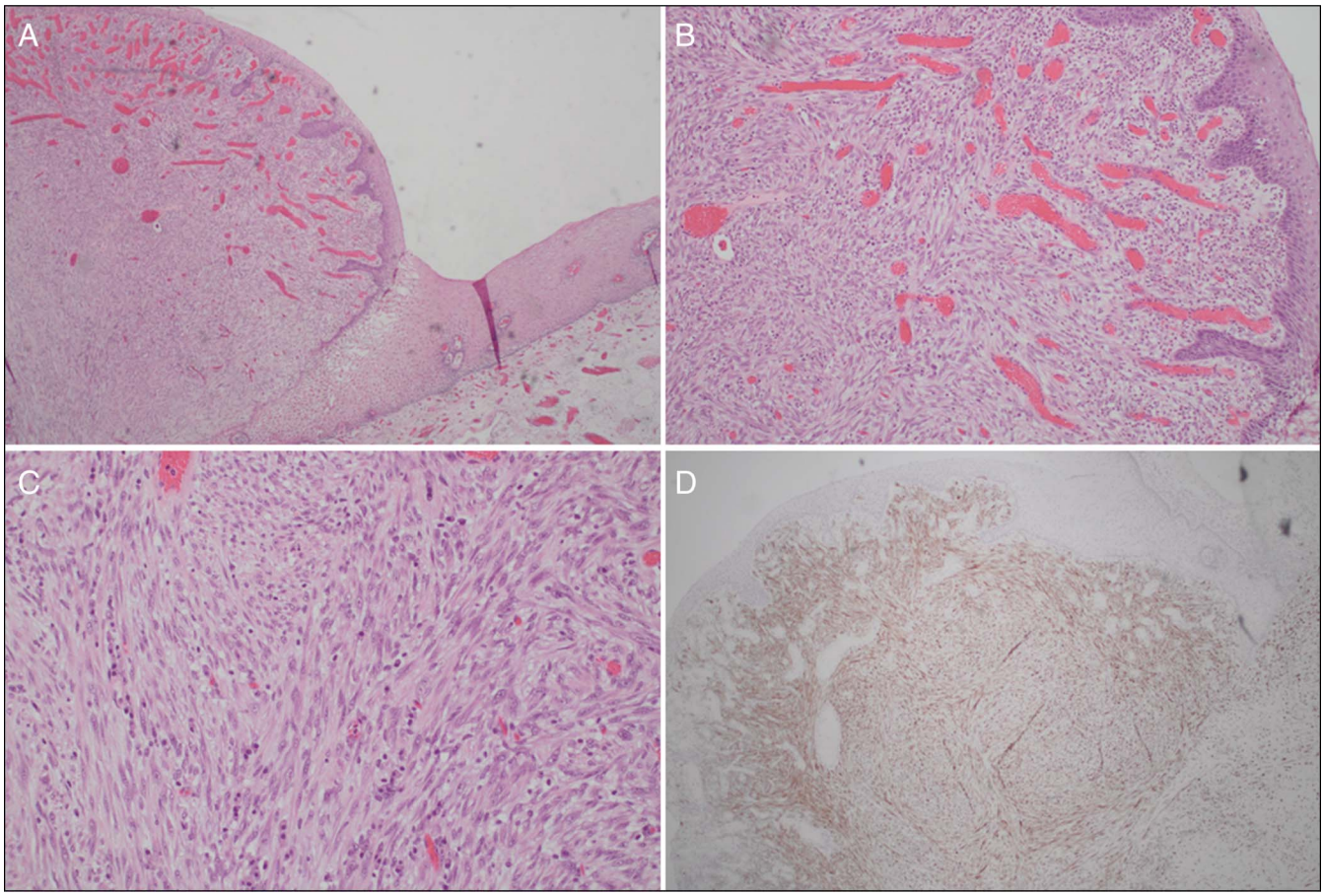

Figure 3. Pathologic evaluation of the tumor: (A) hematoxylin and eosin stain (H&E); 40× magnification. Bland spindle cell proliferation with scattered background inflammatory cells. (B) H&E 100× magnification. Bland spindle cells in vague interlacing fascicles and with scattered background inflammatory cells. (C) H&E 200× magnification. Bland spindle cell proliferation with vague interlacing fascicles and scattered background lymphocytes and plasma cells. (D) Activin receptor-like kinase 1 (ALK-1) stain: Immunohistochemical stain for ALK-1; 40× magnification. Lesional cells stain strongly and diffusely for ALK-1.

(mostly esophagectomy) except for 2 cases; one case was managed with endoscopic mucosal resection using a hot snare in a piecemeal fashion and the second case resolved spontaneously.[5,11] To our knowledge, this is the first case that describes a patient with esophageal IMT that was successfully resected with ESD with no evidence of tumor recurrence after resection at 1-year follow-up. ESD, an organ-preserving procedure, can be considered a valid therapeutic option in early-stage IMTs that spare the muscularis propria particularly in patients who are not surgical candidates and who refuse to undergo surgical resection. ESD is also associated with lower morbidity and better safety profile compared with surgical resection.

DISCLOSURES

Author contributions: V. Muddana performed the endoscopic procedure and was assisted by K. Zakharia. K. Zakharia prepared the manuscript and performed literature review. Manuscription V. Muddana reviewed the final version of the manuscript and is the article guarantor.

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