JCS Journal of Chest Surgery
https://doi.org/10.5090/jcs.20.151
pISSN: 2765-1606 eISSN: 2765-1614
J Chest Surg. 2021;54(6):524-527

Case Report

Pseudomyogenic Hemangioendothelioma Involving the Esophagus: A Case Report

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ARTICLE INFO
Received December 14, 2020
Revised January 20, 2021
Accepted February 9, 2021

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Herein, we describe the case of a 20-year-old woman who presented with dysphagia of 2 months' duration associated with vomiting, moderate abdominal pain, decreased oral intake, and significant weight loss. During the past 3 years, the patient experienced intermittent mild abdominal pain with infrequent vomiting. Endoscopy at Jordan University Hospital showed a mass in the esophagus, and endoscopic biopsies were performed. The preliminary histopathological report excluded malignancy. Two days after endoscopy, the patient presented to the emergency department complaining of severely worsening pain and total dysphagia. The pain persisted despite intravenous paracetamol administration, which was concerning for esophageal perforation; therefore, an urgent surgical intervention was performed. The mass was removed surgically, along with a para-esophageal lymph node. The final histopathological results of the endoscopic and resected specimens supported the diagnosis of pseudomyogenic hemangioendothelioma (PMHE). This is the first case reporting esophageal involvement of PMHE.

Keywords: Hemangioendothelioma, Vascular neoplasms, Deglutition disorders, Esophageal neoplasms, Lymphadenopathy, Case report

Case report

A 20-year-old female patient presented to gastroenterology clinic in Jordan University Hospital, complaining of progressive dysphagia of 2 months' duration, associated with nausea, vomiting, moderate epigastric pain, decreased oral intake, and significant recent unintentional weight loss of 10 kg. The patient's appetite was normal. The physical examination was remarkable for epigastric tenderness and cachexia (weight, 40 kg). During the past 3 years, the patient experienced intermittent mild abdominal pain with infrequent vomiting. She was followed at a private clinic (outside our university hospital), and 2 previous upper endoscopies were done, without proper documentation of the endoscopic or pathologic findings. On both occasions, she was diagnosed with benign distal esophageal ulceration due to gastroesophageal reflux disease and was treated with a proton pump inhibitor with partial improvement. A few days after her visit, endoscopy at Jordan University Hospital showed an obstructive mass, extending from 28 to 33 cm from the incisors. The mass was polypoid, fungating, friable, and ulcerated (Fig. 1A). Multiple endoscopic biopsies were performed. The clinical impression was in favor of a benign mass due to the long duration of symptoms (3 years). A Gastrografin swallow study showed esophageal dilatation, with a large filling defect involving the lower esophagus (Fig. 1B). Abdomen and chest computed tomography showed esophageal luminal obliteration by the mass, diffuse esophageal wall thickening, hiatal hernia with adjacent necrotic para-esophageal lymph nodes (the largest of which measured 8 mm along its short axis), and para-aortic pathologic lymph nodes (Fig. 1C). The bone scan showed no foci of abnormal uptake. The preliminary histopathological report excluded malignancy, but was not conclusive.

Two days later, the patient presented to the emergency department complaining of severely worsening abdominal pain and total dysphagia. The pain persisted despite intra-

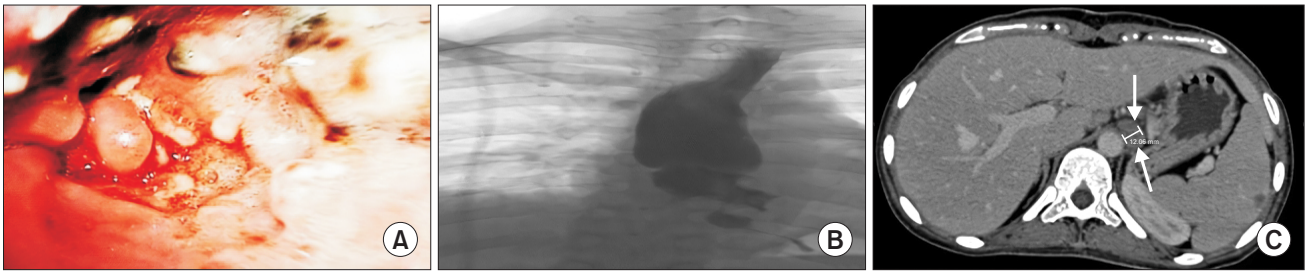

Fig. 1. Imaging of the lesions. Endoscopic examination showing a polypoid, fungating, and friable lesion (A). A Gastrografin swallow study showing esophageal dilatation with a large filling defect involving the lower esophagus above the gastroesophageal junction; a minimal amount of contrast passed to the stomach (B). Axial computed tomography scan showing a para-aortic pathologic lymph node with a size of 12.06 mm (C, arrows).

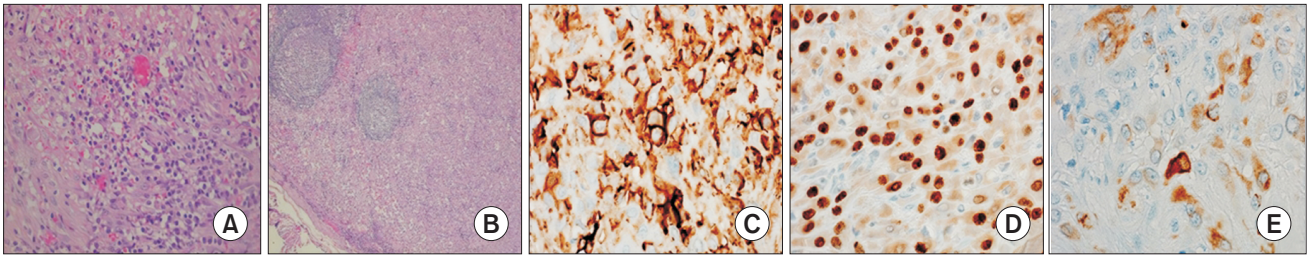

Fig. 2. (A–E) Histopathology of the lesions. Histopathology of the lesions showing neoplastic cells with large eosinophilic cytoplasm and round vesicular nuclei, without significant nuclear atypia. The tumor was associated with an inflammatory background comprised of neutrophils, lymphocytes, and plasma cells (A: hematoxylin and eosin [H&E] stain, ×400). Lymph node involvement by the tumor was evident (B: H&E stain, ×400). Membranous immunoreactivity of the tumor.

venous paracetamol administration, which was concerning for esophageal perforation. Therefore, an urgent surgical resection of the esophageal mass was performed, even without a final histopathological diagnosis. Since the preliminary histopathological report excluded malignancy, we decided to manage the patient with distal esophagectomy without para-aortic lymph node dissection. Distal esophagectomy with gastric pull-through was performed through a left thoracoabdominal incision. The esophageal mass was located 3 cm away from the proximal resection margin, and 2.2 cm away from the distal resection margin. A para-esophageal lymph node around the distal third of the esophagus was also resected.

A few days later, final histopathological results of the endoscopic and resected specimens supported the diagnosis of pseudomyogenic hemangioendothelioma (PMHE). The histopathological examination showed an epithelioid neoplasm with multiple foci of necrosis. The tumor cells were medium-to-large in size with abundant eosinophilic cytoplasm and round vesicular nuclei, without significant nuclear atypia. Few mitotic figures were noted. The tumor cells were infiltrated and surrounded by a polymorphous inflammatory cell infiltrate composed of neutrophils, lymphocytes, and plasma cells (Fig. 2A). Lymph node metastasis of the tumor was also evident (Fig. 2B). Multiple, well-controlled immunohistochemical stains revealed membranous immunoreactivity of the tumor cells for CD31 (Fig. 2C), nuclear positivity of the tumor cells for the vascular marker ERG (Fig. 2D), and focal immunoreactivity of the tumor cells for pan-cytokeratin (Fig. 2E).

The postoperative course was smooth and the patient was discharged on the ninth day following the operation. Six weeks after the operation the patient had gained 8 kg, and did not complain of any recurring symptoms.

The patient provided written informed consent for the publication of her clinical details and images.

Discussion

PMHE was classified by the World Health Organization as an intermediate-grade vascular tumor that rarely metastasizes. The largest series showed that the tumor typically involve the dermis and subcutaneous tissue, with a smaller number involving skeletal muscle and bone [1]. PMHE has a striking male predominance (82%). The mean age at presentation is 31 years, and 94% of patients present during

their second to fifth decades of life [1].

This tumor is extremely rare; to the best of our knowledge and after a careful review of the English-language literature, this is the first case of well-documented PMHE of the esophagus. Furthermore, it is the third reported case describing lymph node involvement of this neoplasm [1,2].

Histologically, the tumor has a nodular architecture, with infiltration toward surrounding adipose or skeletal muscular tissues and an occasional desmoplastic reaction. The tumor cells are arranged in sheets or short fascicles within a background of variably prominent inflammatory infiltrates commonly composed of neutrophils or, less commonly, lymphocytes, plasma cells, or eosinophils. Some cases demonstrate a myxoid background [3].

PMHE seems to have a variable clinical course, with frequent local recurrence, but a small risk of distant metastasis [1].

These tumors are usually treated by surgical excision. Since metastasis is rare, local surgical control is the mainstay of recommended management. All the symptoms related to PMHE are due to local mass effects, as in our case. Since this neoplasm is slow-growing and considered to be of an intermediate grade [3], experts believe that conservative, symptomatic management may be warranted. The few studies investigating this issue demonstrated that some PMHE patients had many years of survival with non-surgical, conservative, and symptomatic treatment [1,4-7].

However, PMHE is known to exhibit local recurrence with the possible development of new lesions; 58% of cases demonstrate local recurrence or the development of additional nodules in the same region, 94.5% of which occur within 1 year of the first presentation [1]. In the event of local recurrence, or the development of new lesions, surgical resection (re-excision) is recommended. In view of the high recurrence rate, regular follow-up is advisable, especially in the first year in which most cases of recurrence take place (94.5%). In addition, long-term follow-up is advisable due to the real, albeit very small, risk of distant metastasis occurring long after the initial diagnosis [1]. Metastatic lesions occur in rare cases [8]. Systemic chemotherapy has been suggested for this group of patients in an attempt to induce tumor shrinkage, and consequently alleviate the tumor mass effects. Unfortunately, substantial clinical trials have not been conducted due to the rarity of PMHE, and different types of systemic chemotherapeutic regimens have been described [8]. Monotherapy regimens included everolimus, sirolimus, telatinib, and gemcitabine. Combined regimens included sirolimus and zoledronic acid, gemcitabine and docetaxel, cisplatin and doxorubicin, cyclophosphamide and prednisolone, and ifosfamide and doxorubicin.

Multifocality, age at presentation, sex, and tumor size may be prognostic factors, but more studies are needed [3]. Generally speaking, long-term survival in affected patients is excellent [9].

Both cases of lymph node involvement previously reported in the literature showed inguinal lymph node and lower limb involvement. The first case involved a 49-year-old woman with a long course of disease and 2 local recurrences. The second recurrence included inguinal lymph node involvement 10 years after excision of the primary subcutaneous tumor. During the course of the disease, the patient underwent 3 surgical procedures (excision of the primary tumor, followed by 2 re-excision procedures for the 2 local recurrences). The second case was an 18-year-old male patient, presenting with PMHE involvement of the thigh, scrotum, penis, and inguinal lymph nodes, all of which were excised surgically. The patient remained disease-free.

Conflict of interest

No potential conflict of interest relevant to this article was reported.

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