



Rare presentation of a gastrointestinal stromal tumor with spontaneous esophageal perforation: A case report

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ABSTRACT

INTRODUCTION: Gastrointestinal stromal tumors (GISTs) of the alimentary canal are malignant tumors with <1% of cases diagnosed in esophagus. These cases require special consideration given their close proximity to vital structures and propensity to be highly aggressive. Management of patients with GISTs has been transformed since the introduction of tyrosine kinase inhibitors. In this report, we present an unusual case of GIST with spontaneous esophageal perforation.

PRESENTATION OF CASE: A 39-year-old Caucasian male presented to our hospital with complaints of severe chest and abdominal pain. A diagnostic chest radiograph revealed a moderate right-sided pleural effusion. Subsequently, an esophagram demonstrated a perforation proximal to an elongated stricture in the distal esophagus. A left thoracotomy was performed whereby a large mediastinal mass firmly attached to the esophagus and gastroesophageal junction was encountered. The neoplasm involved proximal one-third of the stomach and perforated into the right hemithorax. Histopathological evaluation of the tumor led to a diagnosis of GIST.

DISCUSSION: GISTs of the gastroesophageal junction are uncommon and may rarely present with esophageal perforation. The standard of care for treating GIST at present includes tyrosine kinase inhibitors. This pharmacologic agent, along with improved surgical techniques and understanding of molecular markers for accurate diagnosis, will assuredly continue to improve overall survival of patients with GISTs.

CONCLUSION: When stricture or achalasia is detected on imaging, GIST should be considered in the differential diagnosis for individual patients. Additionally, chest and abdomen CT scans of may be performed to confirm presence of a tumor mass, thereby ruling out achalasia.

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1. Introduction

Gastrointestinal stromal tumors (GISTs) are the most common mesenchymal tumors of the alimentary canal that account for 0.1–3% of all gastrointestinal malignancies. Lesions can occur anywhere along the gastrointestinal tract, with the majority arising in the stomach (60%) and small intestine (30%). Primary GIST of the esophagus, however, is rare and constitutes less than 1% of all tumors of its kind.¹ GIST of the gastroesophageal junction presenting with esophageal perforation has seldom been described in the literature. We report here an unusual presentation of GIST with spontaneous esophageal perforation, followed by a

discussion of the unique aspects of prognosis and surgical management of esophageal GIST in comparison to lesions in other primary anatomic sites.

2. Case report

A 39-year-old male presented to our hospital with severe chest and abdominal pain after a few months of progressive dyspnea and odynophagia. Recently, he had complaints of emesis after meals and eventually became intolerant of solids and liquids. Additionally, the patient admitted to an approximately 100 lb weight loss over the past few years. Upon arrival, he was hypotensive and septic, with hemoglobin of 11.9 g/dL and white blood cell count of 18.6 thou/cu mm. After resuscitation and stabilization, a diagnostic chest radiograph revealed a moderate right-sided pleural effusion that was drained via tube thoracostomy with an output of 1200 mL serosanguineous fluid. A subsequent esophagram demonstrated

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a perforation proximal to an elongated stricture in the distal esophagus.

Due to the chronic nature of his symptoms, we made the presumptive diagnosis of achalasia with perforation from a possible food bolus proximally and a surgical intervention was deemed necessary. Our operative plan was to perform a left thoracotomy and if necessary, dissect the diaphragm to mobilize the stomach. A surgical approach from the right chest was decided to be a less ideal by the surgical team due to the difficulties of mobilizing the intra-abdominal esophagus and upper stomach.

A left thoracotomy was performed on this patient, during which a large mediastinal mass firmly attached to the esophagus and gastroesophageal junction was encountered. The visible portion of the mass measured approximately 6 cm and extended down through the diaphragmatic hiatus. During the initial exploration, the esophagus was mobilized with the affixed neoplasm. At this point, the surgical team observed the perforation of the tumor into the patient's right hemithorax. Subsequently, the diaphragm was dissected laterally to expose the intra-abdominal esophagus and stomach. The tumor involved the proximal one-third of the stomach. Since it was impossible to gain distal control of the mass, a midline abdominal exploration was performed followed by a proximal gastrectomy and distal esophagectomy with reconstruction via gastric pull-up and esophago-gastrostomy in the left chest. The diaphragm was then closed around the gastric pull-up. The abdomen and thoracotomy were closed, and chest tubes were placed.

Pathologic analysis of the fibrous neoplasm removed from the patient measured 19 cm × 15 cm × 14 cm and weighed 1291 g (Fig. 1). Histopathological evaluation of the tumor was consistent with a mixed spindle and epithelioid cell GIST. There was necrosis present in approximately 10% of the tumor with a 6 cm break in the pseudocapsule. The mitotic index was 11 per 50 high-power-field. The malignant cells exhibited dense cellularity and marked atypia with a pushing tumor border (Fig. 2). Immunohistochemical studies revealed tumor neoplastic cells that stained positive for KIT (CD117) and CD34, while smooth muscle actin and S-100 protein were negative (Fig. 3).

Postoperatively, the patient initially had an uneventful recovery with no evidence of leak from his anastomosis. He was then started on a diet. Subsequently, he developed an empyema on post-operative day 7 in the left chest requiring an operative decortication from which he recovered completely. The patient did have poor overall fitness for someone of his age. His hospital course was prolonged with an overall length of stay of 15 days due to sepsis and malnutrition.

When the patient had satisfactorily recovered from his hospitalization to tolerate medical treatment, his case was presented at our multidisciplinary tumor conference. The recommendation

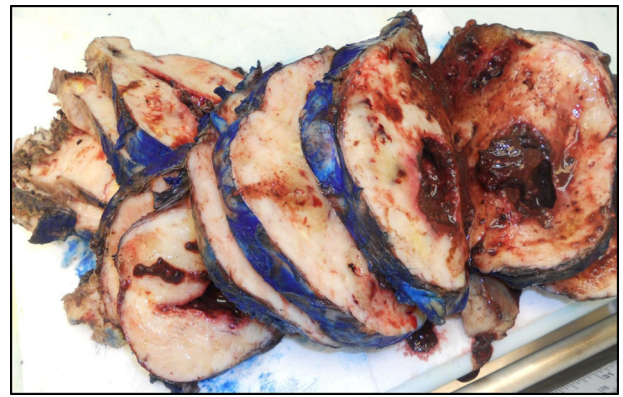


Fig. 1. Malignant gastrointestinal tumor (GIST) of the esophagus, clinically ruptured, gross appearance. This gross photograph of the 19 cm tumor demonstrates the typical fleshy appearance of a GIST. There is an area of cystic degeneration and necrosis, supporting malignancy, which contributed to the clinical rupture of this tumor. The external surface has been marked with blue ink to facilitate evaluation of the tumor.

was made that he receives imatinib for an extended period of time, which would be determined by the status of his overall health and response to the medication.

3. Discussion

Since 1992, there has been a considerable increase in the reported incidence of gastrointestinal stromal tumors. This surge in the number of diagnoses per year may be attributed to greater awareness and improved histopathologic detection of this neoplasm.² It is now believed that GISTs arise from pluripotential mesenchymal stem cells programmed to differentiate into interstitial cells of Cajal in the myenteric plexus.³ A gain-of-function mutation in the juxtamembrane domain of the c-kit gene with constitutive, ligand-independent activation of KIT receptor tyrosine kinase is thought to cause tumorigenesis. Autophosphorylation of the receptor, most often KIT (CD117) or platelet-derived growth factor receptor alpha (PDGFRA), leads to constitutive activation of the receptor and tumor growth.⁴

The immunohistochemical profile of GISTs is characteristic, with nearly 95% of tumors staining positive for KIT (CD117). Often the staining is strongly diffuse in the cell cytoplasm. Less commonly, a membranous or perinuclear pattern is observed. The intensity of KIT staining can be inconsistent. A proper titer of the KIT/CD117 antibody must be used in order to avoid over-staining that could mislabel other neoplasms as GIST. Staining intensity, however, has no predictive value of tumor response to treatment.⁵ KIT-positivity can also be found in other tumors, including melanoma, angiosarcoma and neuroblastoma, and therefore is not sufficient to make

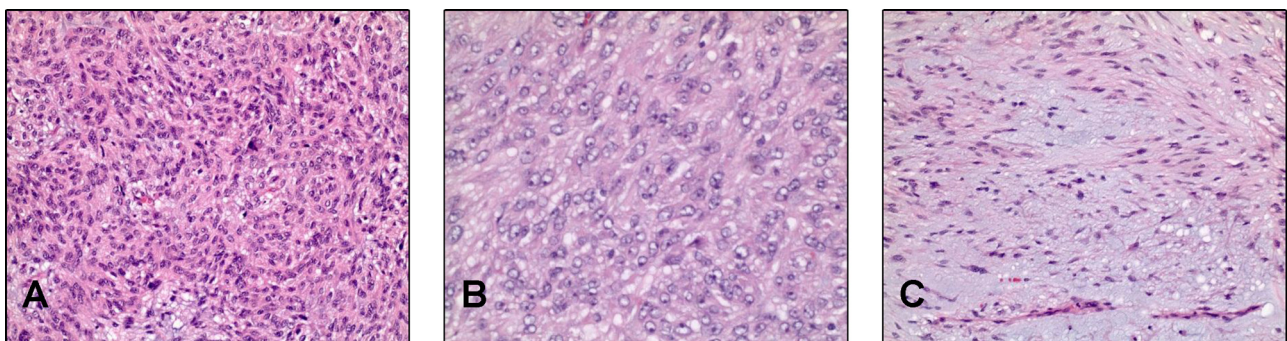


Fig. 2. Malignant gastrointestinal tumor (GIST) of the esophagus, microscopic appearance. The tumor demonstrated (A) overall dense cellularity, a mixture of (B) epithelioid and (C) spindled areas growth, and brisk (11/50 high power fields) mitotic activity with about 10% necrosis (not illustrated), all of which contribute to this tumor being designated as malignant.

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