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Giant 10 cm Diameter Esophageal Gastrointestinal Stromal Tumor (GIST)

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Abstract

Gastrointestinal stromal tumor (GIST) is a relatively rare type of cancer that affects an estimated 5000 to 10,000 new people each year in the US. Most GIST tumors develop in the stomach; but they can also arise in the small intestine, colon and rectum, esophagus or abdominal cavity. Gastrointestinal stromal tumors are the most common mesenchymal, nonepithelial tumors of the gastrointestinal tract in adults between 40 and 50 years of age. Esophageal GIST, in contrast, is rare, amounting to 12.7% - 28% of mesenchymal esophageal tumors or 2% of all GISTs and their diagnosis and management are still challenging, as illustrated in the following case. However here we report a rare case of a 43-year-old female patient who was referred to our department complaining with a 1-month history of atypical chest pain and productive cough. A chest computed tomographic scan and endoscopic ultrasound revealed a submucosal esophageal tumor measuring 100 × 80 × 40 mm in its largest diameter, we performed enucleation via left thoracotomy.

Keywords

GIST, CT Scan, Esophageal Tumors, Gastrointestinal Endoscopy, Subcarinal Lymph, Thoracoscopic

1. Introduction

Until about 20 years ago, GISTs were actually thought to be a totally different type of cancer. The discovery of the molecular switch, called KIT that turns on the growth of GIST has made it possible to diagnose GIST and has led to the development of gastrointestinal stromal tumors GISTs which are the most common mesenchymal neoplasms of the gastrointestinal tract [1]. Consequently, there was interest in these tumors and their management and prognosis has been extensively investigated and standardized. GISTs are rare before the age of 40 years and they are rare in children; we have found that many tumors earlier classified as gastrointestinal smooth muscle tumors in children actually are inflammatory myofibroblastic tumors [2]. Stromal tumors of the gastrointestinal tract represent rela-

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tively rare lesions that are thought to arise from connective tissue elements located along the entire length of the gut. For many years these tumors have been the subject of much controversy and debate in the literature regarding their histogenesis, criteria for diagnosis, prognostic features, and nomenclature. Only a minority of these lesions, mainly those confined to the esophagus and rectum, have been shown to correspond to mature, well-differentiated types of neoplasms such as leiomyoma or leiomyosarcoma of the conventional type. The majority of stromal tumors of the gastrointestinal tract correspond to a heterogeneous group of lesions that have their common denominator as an immature proliferation of epithelioid or spindle cells arising from its muscle layer and showing partial or incomplete myoid, neural, ganglionic, or mixed features of differentiation. GISTs are most common in the stomach (60% - 70%), followed by small intestine (20% - 30%), colon and rectum (5%) and esophagus (<5%). Occasional GISTs primary in the omentum and mesentery have also been reported [3] [4]. The primary site for a malignat GIST extensively involving the abdominal cavity may be impossible to determine.

2. Case Report

A 43-year-old female was referred to the our Cardiothoracic Surgery Department for evaluation of an intramural esophageal mass. The patient complained of atypical chest pain of gradual onset over the previous two months. She denied weight loss, dysphagia, upper GL bleeding, reflux, or other symptoms. The patient's medical history included productive cough. After a chest radiograph failed to show any pathology, a computed tomography (CT) scan was ordered which revealed showed a 100 × 80 × 40 mm mass in direct contact with the esophagus (**Figure 1**). The mass was approached via a left posterolateral thoracostomy. The mass and subcarinal lymph nodes were found to be enlarged and were sent for frozen section, which was negative for malignancy (**Figure 2**).

Opening the mediastinal pleura and manufacture of performs with careful detachment of the muscle layer and the mucosa of the esophagus. The muscular layer of the esophagus was repaired with Vicryl 3/0 sutures. Check for bleeding and air leak from the lung parenchyma. An upper gastrointestinal series on postoperative day 1 showed no evidence of a leak and the patient was uneventfully discharged on the 6th postoperative day. Histopathological evaluation of the tumor led to a diagnosis of GIST

Immunohistochemical testing proved positive for markers CD117, SMA, Desmin and Vimentin but it was negative for CD34, S-100 and GFAP. The cellular proliferation index MIB-1 is 1%. The patient is closely followed up with endoscopy and CT scans every 3 - 4 months for 3 years and is currently free of disease two years after surgery.

3. Discussion

Complete surgical resection is the standard treatment for localized GISTs; however, 40% to 90% of curative resection patients experience recurrence. Esophageal GISTs are rare, highly vascularized tumors [5]. Large tumors specifically have a tendency to rupture or have a risk of a positive margin despite a macroscopically complete

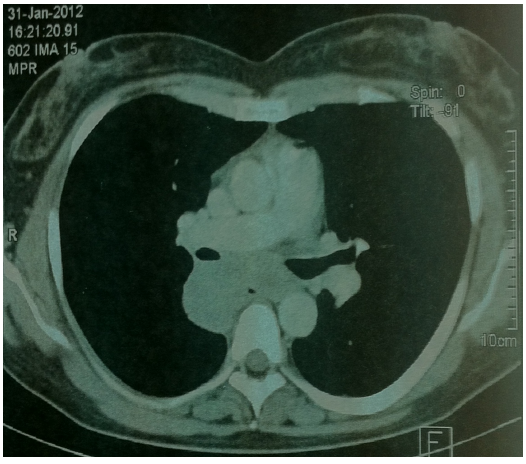

Figure 1. Computerized tomography (CT) scanning of the chest revealed a completely calcified 100 × 80 × 40 mm mass at the right lower post mediastinum.

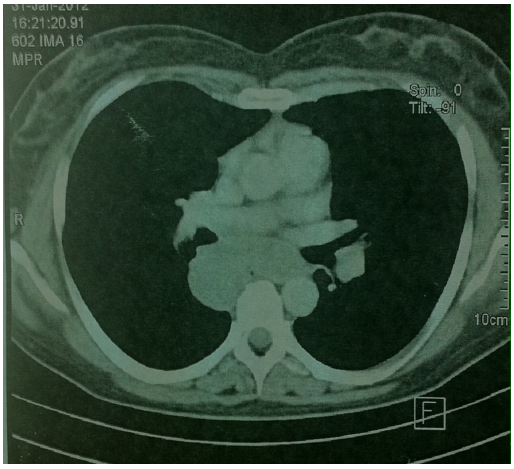

Figure 2. CT scan of the chest, showing a well-circumscribed 10 cm submucosal esophageal mass.

resection. Tumor rupture or the presence of a residual tumor is strongly associated with recurrence and poor prognosis.

A selective approach to biopsy based on tumor size (>5 cm) and suspicious radiological appearance is warranted until the risks of biopsy compared to the benefit of accurate preoperative diagnosis and planning are determined. we did not perform a preoperative biopsy in our patient, while a PET scan might have been appropriate, but it was not possible to obtain it with the patient's insurance in our institution. Each patient should be evaluated individually by surgical risk, tumor size and biology and should actively participate in the management decision process.

Risk factors for gastrointestinal stromal tumor (GIST): Many cases who get the disease may have few or no known risk factors. Currently, there are very few known risk factors for gastrointestinal stromal tumors (GIST): Genetic syndromes, Familial gastrointestinal stromal tumor syndrome, Neurofibromatosis type 1, Carney-Stratakis syndrome.

Traditional histologic criteria are not specific enough to diagnose GIST. This diagnosis must be confirmed with CD117 Immunohistochemical staining.

4. Conclusion

Esophageal GIST is a rare disease and complete surgical resection is the standard treatment. Regular follow-up is needed even if resection is complete and negative margins are achieved. In this paper we have reviewed current knowledge of gastrointestinal stromal tumor (GIST) management. Surgical resection is the treatment of choice for these tumors. GIST studies have shown that after surgical resection these tumors span a wide clinical spectrum from benign to malignat tumors. While low malignat potential GISTs have an excellent GIST involving a high rate of recurrence with poor survival after surgical treatment alone.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images.

Conflicts of interest

1) The material has not been previously published or submitted elsewhere for publication; 2) There are not conflicts of interest of any of the authors.

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