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A Case of Giant Esophageal Gastrointestinal Stromal Tumor that was Successfully Resected without Neoadjuvant Treatment

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Received Date: 29-01-2022; Accepted Date: 24-02-2022; Published Date: 03-03-2022

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Abstract

A 27-year-old male from Gaza was admitted to the Gastroenterology department with complaints of progressive dysphagia. An endoscopic examination shows submucosal gastric mass at the Gastro-Esophageal (GE) junction. Physical examination was normal. There was no abnormality found in laboratory examinations, including hematologic and biochemical analyses. A whole abdominal enhanced Computed Tomography (CT) scan revealed a solid mass with a smooth border and hypervascularity at the gastro-esophageal junction. Endoscopic ultrasound guided fine needle biopsy was performed, pathologic diagnosis of the submucosal tumor was GIST. The patient was considered for neoadjuvant immunotherapy, but then he is excluded due to logistic considerations.

The patient underwent distal esophagectomy and proximal gastrectomy, complete resection was performed and the specimen was sent for histopathology. The diagnosis of an esophageal GIST with negative margins was confirmed by histological investigation of the resected specimen. Spindle-shaped tumor cells were discovered in the tumor. CD34 and DOG1 were shown to be positive in the tumor cells by immunohistochemistry. In 50 high-power fields, the

Hemedat B | Volume 3; Issue 1 (2022) | JSRP-3(1)-033 | Case Report

Citation: Asbeh YA, et al. A Case of Giant Esophageal Gastrointestinal Stromal Tumor that was Successfully Resected without Neoadjuvant Treatment. Surg Res Prac. 2022;3(1):1-7.

DOI: <http://dx.doi.org/10.46889/JSRP.2022.3104>

mitotic index was less than 5 mitotic figures. There were no postoperative complications. The patient was discharged 5 days after the surgery and has been doing well since then. A repeat endoscopy upper GI and a CT scan of the whole abdomen will be scheduled during the next follow-up visit to exclude any kind of recurrence or metastases.

We present this unusual case to show the potential of surgical resection in patients with large esophageal GISTs without preoperative imatinib treatment, especially when the patient is unable to receive neoadjuvant treatment for some reason, as long as we can achieve complete resection without rupture and with a clear margin.

Keywords

Case Report; Giant; Esophageal; GIST; Gastrointestinal Stromal Tumor; Neoadjuvant Treatment

Introduction

GISTs (Gastrointestinal Stromal Tumors) were originally detected in the 1980s but did not become widely recognized until the twenty-first century [1]. GISTs are the most frequent mesenchymal neoplasm of the Gastrointestinal tract (GI), representing about 1 to 2% of all primary GI malignancies, with a typical diagnosis age of 65 to 69 years [2,3].

Esophageal GISTs are extremely rare, accounting for less than 1% of all primary GISTs in the gastrointestinal tract.

It is most likely to develop from the Muscularis propria and arise from the interstitial cells of Cajal (ICCs) [4]. GIST positive for c-KIT (CD117) or platelet-derived growth factor PDGFRA gene, DOG1, CD34 and other genes in 90% of cases [5,6].

We presented a patient with a large mesenchymal tumor in the esophagus with the pathognomonic feature of GIST that was successfully treated without neoadjuvant chemotherapy.

Case Presentation

A 27-year-old male patient from Gaza Strip has been admitted into our hospital with dysphagia, regurgitation and epigastric pain. Gradually, he became intolerant to solid and liquid food and he had been losing body weight day by day. He had no significant family history of suffering from such a type of complaint. He had no history of corrosive ingestion. He is a non-smoker. No history of alcohol or drug abuse. The general physical examination was unrevealing. An

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abdominal examination revealed mild epigastric tenderness, with otherwise unremarkable findings. Laboratory tests, including hematologic and biochemical analyses, revealed no abnormalities. Upper endoscopy revealed a large submucosal mass at the gastroesophageal junction, multiple erosion at the first and second parts of the duodenum (Fig. 1). Endoscopic ultrasound revealed a large ulcerated submucosal mass (Fig. 1). The *Helicobacter pylori* test was strongly positive. Endoscopic ultrasound showed a large submucosal esophageal mass extending to the gastroesophageal junction, arising from the muscularis propria.

After an endoscopic ultrasound-guided fine-needle aspiration biopsy, the tumor was diagnosed as a GIST, characterized pathologically by H and E staining and immunostaining. A Computed Tomography (CT) scan showed a mass, $10 \times 9 \times 4$ cm in diameter (Fig. 2), at the lower esophagus and the maximum Standardized Uptake Value (SUV max) was 5.9 on a Positron Emission Tomography (PET)-CT scan (Fig. 2). There was no apparent distant metastasis.

Our clinical diagnosis was GIST at the lower end of the esophagus involving the gastroesophageal junction. We formed a multidisciplinary team composed of general and thoracic surgeons, anesthesiologists, pathologists, radiologists and oncologists. Our plan was for neoadjuvant and surgical resection. However, due to the occupation's policies on dealing with referrals of patients coming from Gaza to the West Bank, there is procrastination and a fear that the patient will not be allowed to return to the hospital. So instead, surgical resection was only performed, IVOR-LEWIS esophagectomy (right thoracotomy and upper abdominal laparotomy, resection of the distal esophagus and proximal stomach, anastomosis and jejunostomy).

After opening the abdomen, the opening of the gastrohepatic ligament was done and we found a firm to almost solid globular mass in the lower end of the esophagus extending up to the fundus of the stomach. Macroscopically, the appearance of the tumor was an encapsulated tumor of a heterogeneous consistency, with alternating areas of necrosis and fibrous strictures. We dissected around the stomach, closing its vasculature and keeping the right gastroepiploic artery as the artery supplying the conduit, D2 lymph node dissection was performed, feeding jejunostomy tube was performed, the closure of the abdomen and moving to right thoracotomy in the 6th intercostal space, dissecting around the esophagus, reaching above the level of the azygous arch and down to the diaphragmatic curare, identification of the thoracic duct, with taking care not to injure it, sampling of mediastinal LN station 7, 8 and 9R. Performing proximal gastrectomy and distal esophagectomy using the endoscopic staplers, then performing the anastomosis, in the chest. A chest tube was inserted, operative and postoperative courses were uneventful.

Pathological evaluation of the resectable specimen relieved a 10 cm x 9 cm x 3.8 cm tumor mass of the spindle cell type (Fig. 3). All surgical margins are negative. The immunohistochemistry for CD34 and DOG1 was positive (Fig. 3). He was discharged home

on a postoperative Day Five and subsequently received adjuvant imatinib therapy (400 mg/day).

The patient was followed up with phone calls and video calls initially as he is unable to come back to the West Bank again. He is doing very well clinically with no dysphagia or abdominal pain. He is on Imatinib therapy at 400 mg/day with no significant side effects to the treatment.

Figure 1: Preoperative image. (A) Endoscopic ultrasound revealed a large ulcerated submucosal mass. (B) Endoscopic image showing the tumor with a smooth surface in the lower esophagus.

Figure 2: CT and PET-CT images. (A) CT scan. The tumor is 10×9×4 cm in diameter. (B) PET-CT scan. SUV max was 5.9.

Figure 3: Resected surgical specimen. (A) The resected tumor was approximately 10 cm x 9 cm x 3.8 cm in diameter. (B) High power view, H&E stain The tumor is hypocellular and composed of spindle cells with eosinophilic cytoplasm, in a collagenous stroma. (C) Intermediate power view, H&E stain the tumor cells are arranged in patternless pattern. (D) DOG1 immunohistochemical stain: The tumor cells show diffuse reactivity to DOG1 immunostain.

Discussion

Our patient presented with an esophageal mass with a pathognomonic feature for GIST, in a setting of progressive dysphagia and weight loss. GISTs are known to arise from mesenchymal tissue of the gastrointestinal tract and specifically arise from the Interstitial Cells of Cajal (ICCs) and develop from the Muscularis propria. From existing epidemiological data we face GISTs are rare in the esophagus with an incidence of less than 1%. GISTs are known to be sporadic tumors [1,2]. They have been linked to syndromes such NF-1, Carney triad (pulmonary chondroma, gastrointestinal stromal tumors and paraganglioma) and familial gastrointestinal stromal tumors in some cases [7].

The pathologic, cytologic and immunohistochemical studies of tumor cells are used to establish the diagnosis. The tumor cells were diffusely positive for CD34 and DOG1 in both smear

specimens and paraffin sections using immunohistochemistry. Most GISTs, including the one in our patient, have a spindle cell morphological origin.

Since the risk of malignancy is increasing with an increase in GISTs size [8]. In the case of 10 cm GISTs, imatinib is used preoperatively as neoadjuvant therapy to help shrink the tumor size and then complete surgical resections are the standard treatment for localized GISTs [9]. However, our patient did not have a chance to get his imatinib mesylate due to occupational obstacles and we were able to perform surgical resection without collateral damage during the surgery and with a negative pathological margin.

Although laparoscopic resection is newly used, in this case, it is not suitable because of the huge size of the tumor and the risk of capsular injury [9].

In multiple clinical investigations, adjuvant imatinib therapy after GIST resection has been found to reduce recurrences and prolong survival. Our patient was discharged on an imatinib dose of 400 mg daily for a one-year duration. He will be followed up by upper GI endoscopy every six months and chest and abdominal CT scan every six months for five years.

In summary, we described a case of a large esophageal GIST that was effectively removed without the use of imatinib, which is a challenging case to treat without neoadjuvant therapy. This was due to the occupation's situation in gaining access to our patient, which set this case different from others.

Conclusion

GIST should be kept as a differential in the evaluation of any submucosal mass arising from the gastrointestinal tract, we treated a rare case of a large esophageal GIST that was successfully resected without neoadjuvant imatinib treatment due to occupational obstacles, with the same surgical outcome compared to other cases treated with neoadjuvant.

Ethical Consideration

The patient agreed to participate in the presentation and publication of the case report with accompanying images.

Conflict of Interest

It is stated that there are no conflicts of interest.

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