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Giant GIST tumour successfully resected in 72 year old Ugandan male

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Abstract

Gastrointestinal stromal tumors (GIST) account for ≤1% of all GIT tumors. They arise from the interstitial cells of Cajal within the myenteric plexus. The gastric GIST can grow to a massive size and a patient will usually present with nonspecific symptoms like abdominal pain and early satiety. Diagnosis is usually a dilemma. We present a 72 year old male who presented with a complaint of abdominal pain for ten years. Imaging studies were not conclusive on type of tumour. Due to early symptoms of gastric outlet obstruction we performed surgery. The tumour was locally advanced and measured 18 cm X 12 cm X 24 cm. Haemoxylin-eosin staining showed spindle cells in whorls while immunohistochemistry was positive for c-KIT (CD117) and CD -34. A diagnosis of GIST was made. Neoadjuvant imatinib was scheduled for the patient.

Keywords

GIST; abdominal pain; diagnosis; tumour.

Introduction

GISTs are uncommon GI tumors accounting for 1% to 3% [1]. They are, however, the most common mesenchymai neoplasms of the gastrointestinal tract [2]. GISTs arise from mesenchymal stem cells which are programmed to develop into the interstitial cells of Cajal located in the myenteric plexus [3]. The cells of Cajal form a complex network within the walls of the GIT and act as a pacemaker [4]. Hence, the GIST tumour can be found anywhere along the GIT. Most frequent presentation is the stomach (60%), small intestine (30%) and colon, esophagus, rectum and anus (10%). GISTs are a result of mutation of KIT and Platelett Derived Growth Factor Alpha (PDGFRA) [5].

GIST tumors cause no characteristic symptoms and thus preoperative diagnosis is defied or delayed. In addition, the tumour can grow to a massive size [1]. Patients will present with delayed symptoms like

abdominal pain, palpable mass, early satiety or anemia due to digestive bleeding. A precise preoperative diagnosis is often a challenge in giant GIST tumors. Surgical resection is the mainstay of non-metastatic disease; Neoadjuvant imatinib mesylate is used for locally advanced disease and has long term positive results [6].

In this report, the diagnostic challenge of a giant gastric GIST tumour is presented.

Case Report

A 72 year old male was referred to Uganda Martyrs hospital, Lubaga, by his local doctor with a chief complaint of abdominal pain for 10 months. The pain was associated with early satiety and weight loss. On physical exam, there was gross distension of his abdomen with a hard, non-tender mass in the left upper quadrant. Laboratory results showed moderate anemia (Hb 10.1g/dl). Liver and renal function tests were normal. Abdominal ultrasound showed an ill-defined echo complex mass in the left abdominal quadrant most likely of splenic origin.

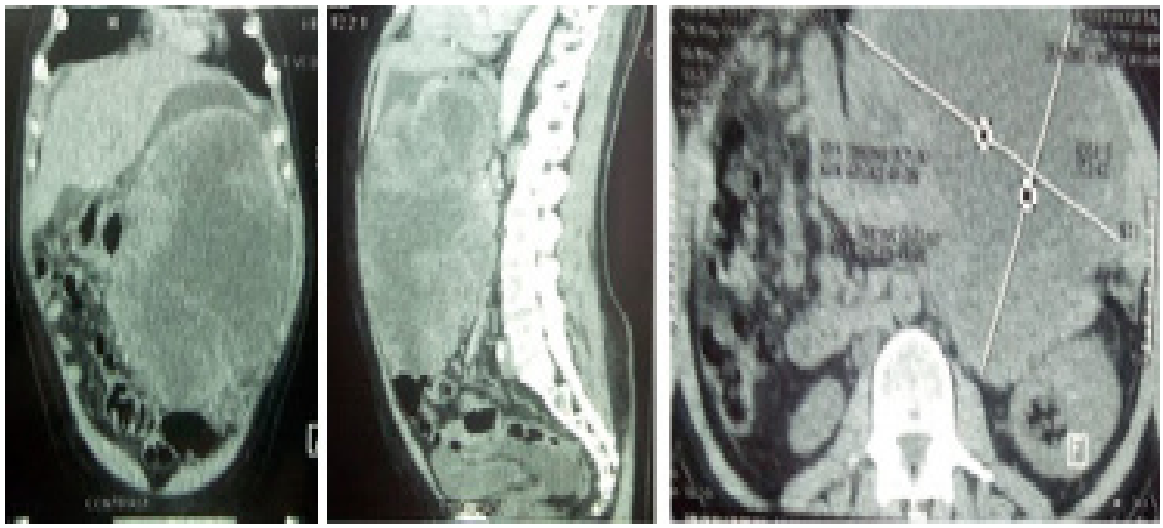

Figure 1: Contrast enhanced coronal, sagittal and axial CT images showing a giant tumour in the abdomen.

Contrasted CT scan showed a mixed density retroperitoneal mass measuring 18 cm X 12 cm. Tru cut biopsy showed morphologic features reminiscent of a hemangioma. Our preoperative diagnosis was a massive spleen and we planned to do splenectomy. Immunological tests of antibodies to HIV and, Hepatitis B were negative.

At surgery, a massive left upper quadrant cystic, septated, retroperitoneal tumour infiltrating and involving the spleen, greater curvature of stomach and transverse colon was found. The tumour was mobilized from its retroperitoneal attachments, the splenic vessels were ligated and the entire tumour was removed piecemeal. Regional lymphadenectomy was not done in view of patients significant blood loss, hypotensive state and transfusion need.

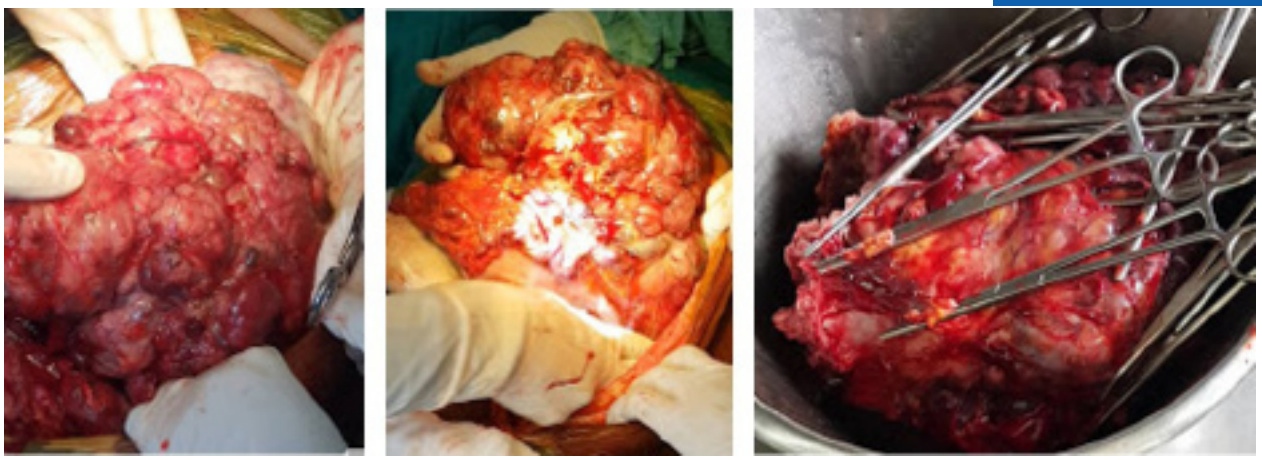

Figure 2: Highlights of intraoperative and postoperative images. Grossly, was a hemorrhagic, heterogeneous solid mass with cystic degeneration measuring 18 cm X 12 cm X 24 cm.

A sleeve gastrectomy of the involved part was done and a transverse double bowel colostomy fashioned. Histology showed atypical pleomorphic spindle shaped cells with abnormal mitoses in a collagenous stroma involving the greater curvature of stomach. Immunohistochemistry showed positivity for CD 34 and CKIT. A final diagnosis of GIST was made. The patient was discharged after 10 days. Patient returned 8 weeks later for a planned colostomy reversal which was successfully done. Patient is scheduled to start chemotherapy with imatinib mesylate.

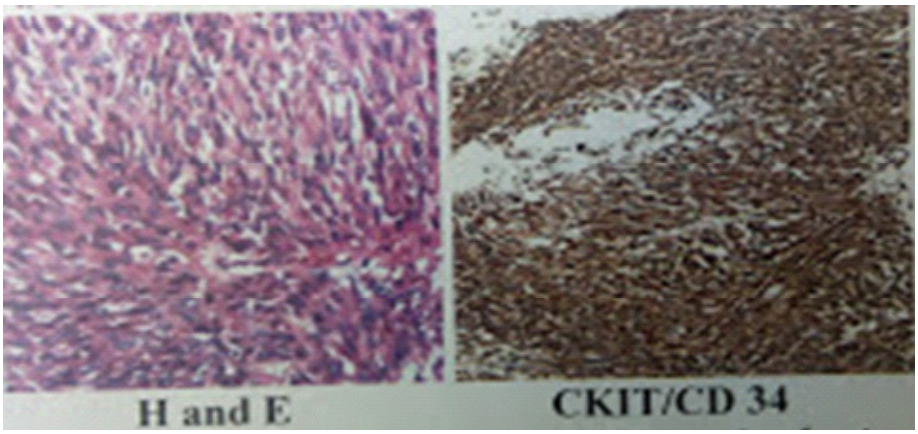

Figure 3: Haemoxylin-eosin staining showing evidence of growth of spindle cells. Immunohistochemical staining was positive for KIT (CD117) and CD-34.

Discussion

Giant GISTs of the stomach are considered to be 10 cm and above in diameter [7,1]. One of the challenges met in management of this patient was getting an accurate preoperative diagnosis. Initially, it was thought to be a large spleen. Due to its asymptomatic nature, the gastric GIST can reach considerable sizes. The largest recorded GIST tumour in the literature so far is 42 cm X 31cm X 23 cm [8]. In this case, the tumour was 18 cm X 12 cm X 24cm. The tumour was locally advanced and neoadjuvant imatinib mesylate to shrink the tumour would have been the ideal line of management. The diagnostic challenge, the complex nature, size of the tumour and the early gastrointestinal obstruction as signaled by patients early satiety made us decide on surgical intervention for the patient.

In addition, identification of the primary site of the tumour was also a challenge. Early satiety was a late feature. Literature reports that 60% to 70% of the GIST tumors are found in the body or fundus of the stomach [5]. Matsuo et al reported the use of 3D CT angiography to help detect primary site of the tumour which helped in planning a safe operation [7]. The tumour in this study had cystic changes which are a uncommon presentation of GIST. Cystic changes are usually suggestive of malignancy because GIST tumors are typically solid organs [9].

Prognostic factors used to determine outcome and risk for recurrence include tumour size and mitotic count [10]. GIST arises from the c-KIT (CD 117) positive intestinal cells of Cajal. Histologically, the tumour has interlacing bundles of spindle shaped cells [11]. Conclusive diagnosis depends on immunohistochemistry. CD117 is positive in 90% of cases, DOG 1 is positive in 95% of cases and CD34 is positive in 50 to 80% of cases [12]. It is paramount to distinguish GIST from other mesenchymal tumors as they are resistant to conventional chemotherapy and radiotherapy [13].

The patient will receive adjuvant imatinib mesylate. Imatinib mesylate is recommended in management of advanced GIST [14]. The ACOSOG clinical trial reported long term recurrence free survival of patients with tumour greater than 3 cm. Following this, imatinib was approved for use as adjuvant treatment of GIST in the USA and Europe [15].

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