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A RARE CASE REPORT: JEJUNAL GASTROINTESTINAL STROMAL TUMOR

Surgery

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ABSTRACT

Gastrointestinal stromal tumors (GISTs) are rare tumors. GISTs account for only 0.2% of all gastrointestinal tumors and only 0.04% of small intestinal tumors. Jejunal GISTs are the rarest sutype. Out of them only 10-30% turn malignant [3]. We present a case of a 60 year old female, who presented with a gradually increasing mass in the umbilical region since 6 months and recent abdominal pain and vomiting. This case report highlights the rarity of jejunal GISTs and their presentation as abdominal lump. In patients with a virgin abdomen ,underlying tumors should be considered.

KEYWORDS

INTRODUCTION:

Gastrointestinal stromal tumours (GISTs), first described by Mazur and Clark in 1983, are rare mesenchymal tumours of the alimentary tract [1]. They are believed to result from activating mutations of proto-oncogenes c-KIT or platelet-derived growth factor receptor alpha polypeptide. These mutations increase tyrosine kinase receptor activity, resulting in uncontrolled proliferation of stem cells that differentiate into intestinal cells of Cajal [2]. The vast majority of GISTs occur in a sporadic and isolated form, but can be features of multiple neoplastic syndromes. GISTs comprise only 0.2% of gastrointestinal (GI) tumours and are mainly gastric tumours. Only 20% of GISTs, i.e. 0.04% of all GI tumours, are small intestinal GISTs and jejunal GISTs are the rarest subtype. Only 10–30% progress to malignancy [3].

CASE REPORT:

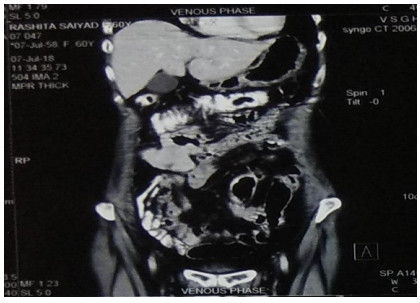
A 60 year old female presented with complaint of gradually increasing mass in the umbilical region since 6 months associated with vomiting and fever since 15 days. Blood tests were normal.

She had no significant past medical or surgical history.

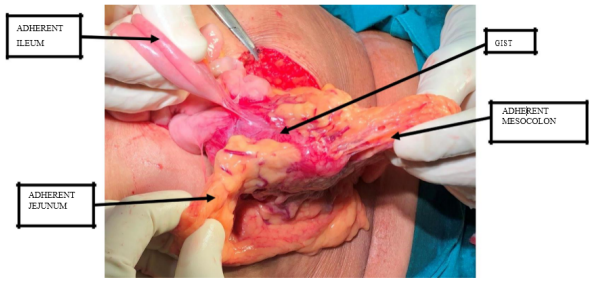
On examination, abdomen was soft, non-distended with a palpable lump of size 5x4cm in the umbilical region, dull on percussion.

USG showed a 65x35mm sized exophytic lobulated mixed echogenic lesion with vascularity arising from the bowel and thickening of bowel wall measuring 11mm.

CECT abdomen revealed a 70 x 53 x 41 mm sized heterogenously enhancing well defined lesion with central non-enhancing necrotic areas arising from mid jejunal loop suggestive of GIST.

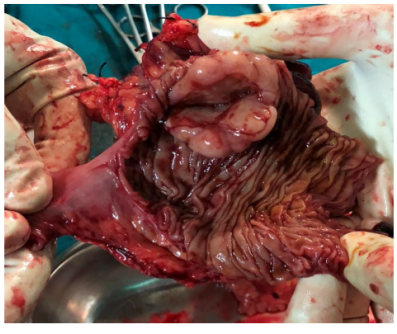


Exploratory laparotomy was performed and approx 6x4cm sized mass was found arising from jejunum approximately 10 cm from DJ flexure with adherent mid jejunal and distal ileal loops. Adhesiolysis was done.



Resection of the 80cm segment of jejunum starting from 5cm distal to DJ flexure and containing the mass and the adherent jejunal loop was done and jejuno-jejunal anastomosis was done.Approximately 10 cm segment of the distal ileal loop, with 5cm clear margins, which was adherent to the mass was resected and ileo-ileal anastomosis was done.

Biopsy report revealed GIST with moderate risk of progressive disease. Stage-II $T_3N_0M_X$ (MI LOW) with tumor free resected margins.



The patient recovered well postoperatively. Staging positron emission tomography (PET)–CT scan after 3 months showed no evidence of recurrence. The patient had postoperative imatinib for 2 years, as per the NICE guidelines for fully resectable GISTs over 10 mm.

DISCUSSION:

Over 90% of GISTs occur in adults over 40 years old. The incidence peak of diagnosis is 60–65 years. There is a slight male predominance but no association with geographic location or ethnicity [2].

Jejunal GISTs are typically asymptomatic while small and may be diagnosed incidentally from CT, endoscopy, during surgery or from symptomatic liver metastases. Enlargement causes variable symptomatology; GI bleeding or non-specific GI symptoms such as

bloating or early satiety. Around 40% are associated with ulceration, and 28% presenting with overt GI bleeding. Bleeding may be acute (haematemesis or malaena) or chronic (anaemia). Around 20% grow large enough to present with pain, a palpable mass or obstruction secondary to intussusception [4].

Barium studies identify 80% of GISTs, capsule endoscopy 81.1%, CT scans 87% and MRI scans close to 100% [3]. Certain factors make diagnosis challenging. Exophytic growth with minimal or no luminal protrusion, which is common, makes endoscopic diagnosis difficult. Poor bowel filling and necrotic areas make GISTs difficult to visualize on CT and cyst degeneration may be misdiagnosed as abscesses or inflamed intestinal loops [5].

Immunohistological and pathological tests are diagnostic when results are combined. Immunohistochemical assay for CD117 antigen, an epitope of the KIT receptor tyrosine kinase, is the mainstay of diagnosis. Approximately 95% are positive for CD117 antigens. However, false-positive results may occur due to weak reactivity to other mesenchymal neoplasms. The morphology of jejunal GISTs is varied: tumours may be composed of spindle cells (70%), epithelioid cells (20%) or mixed spindle and epithelioid cells (10%). Similar histological features may be seen with leiomyosarcomas and leiomyoblastomas. Definite diagnosis therefore relies on a combination of both immunohistochemical assay and morphological histology [6].

Current recommendations for assessing the risk of compression rely on three parameters: tumour size, location and mitotic index. In the presented case, a 4-cm maximal diameter jejunal GIST with one mitosis per high power (5 mm) field indicates a low-to-intermediate probability of metastases and 40–60% of recurrence risk [7].

Surgical resection is the primary treatment for jejunal GISTs. Evidence does not indicate an optimal resection margin size, but a negative margin is vital to prevent local recurrence. Lymph nodes are rarely involved and as such their dissection is not typically indicated. Complete resection of a low-to-intermediate risk GIST, such as the presented case, results in 95% 5-year survival. For GISTs exceeding 10 mm, the National Cancer Institute recommends adjuvant imatinib. This is currently the only effective drug for GISTs. Imatinib gives a 14% absolute reduction in recurrence rate, achieving 97% recurrence-free survival [8].

This case is remarkable for several reasons. First, jejunal GISTs are exceptionally rare, comprising under 0.04% of GI tumours [9]. Secondly, extensive initial investigation which would be expected to be diagnostic yielded false-negative results, resulting in progression to an unusually large jejunal GIST. Thirdly, the patient subsequently presented atypically with mass in abdomen in a virgin abdomen, raising the suspicion of GI tumour [10]. Finally, a clear resection margin was achieved and the patient made a remarkable recovery despite multiple comorbidities.

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