CASE REPORT

Multiple Synchronous Gastrointestinal Stromal Tumors

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ABSTRACT

Gastrointestinal stromal tumors (GISTs) are rare tumors, even more rare is to find multiple, synchronous GISTs in small bowel. We report a case of multiple synchronous GISTs presented as acute intestinal obstruction. Per operatively four tumors were identified in jejunum and ileum with mesenteric lymph nodes involvement.

Key words *Tumor, Stromal, Synchronous, Intestine, obstruction.*

INTRODUCTION:

Gastrointestinal stromal tumors constitute the most frequent group of mesenchymal tumors in the gastrointestinal tract, arising from Cajal's cell.[1,2] Until recently they were classified as being of smooth muscle origin but with the increasing use of immunohistochemistry and their characteristic positive staining for c-kit (CD 117) and CD 34 and negativity for desmin and S 100- protein, have made them a separate entity.[3,4]

GISTs have a wide spectrum in their clinical behavior from benign to highly malignant one. Published evidences suggest that almost all GISTs presenting with clinical symptoms and signs leading to treatment, have potential to behave in a malignant fashion.[3] Commonest site of occurrence is stomach up to 60%, 25-40% arise in the small bowel and less than 10% in the colorectum.[5,6]

This is a case of multiple synchronous GISTs in small bowel who presented as acute intestinal obstruction.

CASE REPORT:

A 59 years old male presented with four day history of pain and distension of upper abdomen associated with vomiting. Clinical examination revealed anemia and tachycardia but there was no fever or signs of dehydration. Abdomen was distended (mainly in upper part), non tender and resonant on percussion. Bowel sounds were audible. Digital rectal examination was unremarkable.

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Base line investigations showed hemoglobin 9.3 Gm/dl and white cell count 10,900/mm^3 with neutrophil count 83%. Serum electrolytes, blood sugar and urine analysis were normal. X-ray abdomen showed distended small bowel loops and multiple air-fluid levels. Initially he was given a conservative treatment (drip and suck) and active observation. In next 24 hours abdominal distension increased with tenderness in upper abdomen. Repeat x-ray abdomen showed no improvement hence, laparotomy was performed.

At laparotomy four circumferential growths were found in the small bowel up to ileo-caecal junction. First growth was approximately 25 centimeters away from duodenojejunal flexure. It was about to give way due to weakening of bowel wall and some degree of necrosis was present over serosa (fig. I). Second growth was present 30 cms distal to the first lesion. It was causing complete obstruction with proximal bowel dilatation (fig. II). Third lesion was found approximately 20 cms proximal to ileo-caecal junction (fig. III) and fourth lesion was at ileo-caecal junction (fig. IV). Multiple enlarged lymph nodes were present in adjoining mesentry and liver was normal. Proximal two growths were resected separately with adjoining mesentry and two end to end anastomoses were performed. Third and fourth growths were resected as right hemicolectomy. Postoperative recovery was uneventful except minor wound infection. CT scan was done postoperatively, which showed normal liver and no evidence of abdominal or pelvic lymphadenopathy. Histopathology was reported as poorly differentiated carcinoma. Immuno-histochemistry was requested and confirmed the nature of tumor as GIST.

DISCUSSION:

Gastrointestinal stromal tumors are rare tumors that account for less than 3% of all gastrointestinal

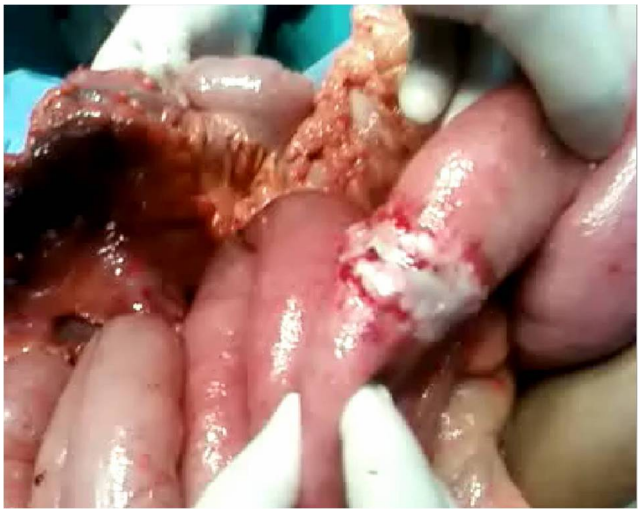

Figure I: Tumor caused weakening of bowel wall showing some degree of necrosis over serosa

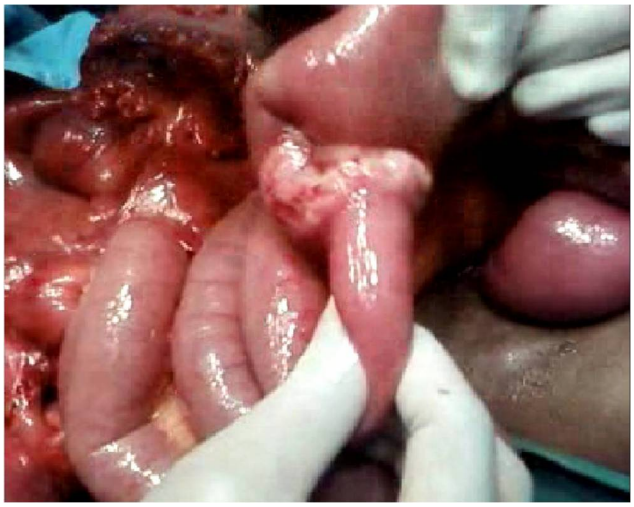

Figure II: This tumor was the cause of complete obstruction with proximal bowel dilatation.

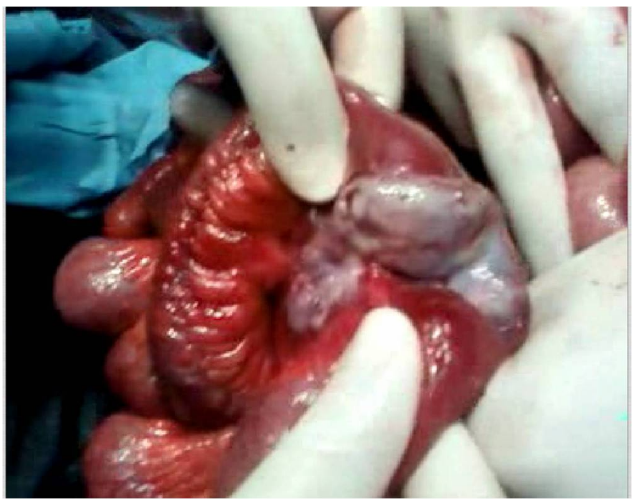

Figure III: This tumor was 20 cm proximal to ileo cecal junction

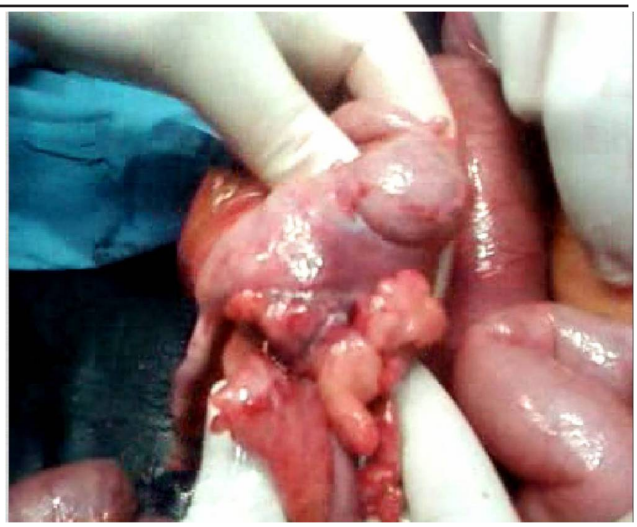

Figure IV: This tumor was present at ileocaecal

neoplasms.[7] The precise incidence of GIST remain unknown as the diagnosis has been modified recently, but is estimated at approximately four per million per year.[8] Peak incidence occurs in patients older than 50 years. Usual presentations are melena, hematemesis, abdominal pain, weakness, rarely weight loss. Acute presentation of intestinal obstruction or perforation can occur.[4,8] Surgical excision is the treatment of choice.[3,9] In past, GISTs were notoriously unresponsive to chemotherapy and radiotherapy and there was no effective treatment available for highly malignant form, tumors with distant metastasis, recurrent tumors or irresectable tumors.[3,4] Now a days imatinib mesylate is in practice for such kind of lesions, because 90% of malignant lesions harbor a mutation in c-kit, which is specifically inhibited by this drug.[3,10]

Literature search showed that various malignant and non-malignant conditions co-exist with the GIST. Chen et al from Taiwan reported a case of triple synchronous tumors in a patient (adenocarcinoma of stomach, adenocarcinoma of gall bladder with stromal tumor of the stomach).[11] Khan et al reported a case of simultaneous occurrence of GIST of stomach with gastric tuberculosis.[7]

It is a rarity to find multiple synchronous GIST simultaneously which was found in index case. An extensive search of published literature revealed only one patient of multiple GISTs in a patient with neurofibramatosis type I (NF1).[2] Retrospectively, keeping in mind NF 1, patient was reviewed but no evidence of this disease was found.

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