laparotomy in those presenting acutely, as in our patient. Melena occurs in 20%-35% of patients\textsuperscript{3,4} but perforation is uncommon.\textsuperscript{1,5}

The bowel perforation may occur in the duplication itself, or very rarely in the adjacent normal intestine that is exposed to acid secreted by the ectopic gastric mucosa. In saccular duplications perforation may be due to the pressure of accumulating intestinal secretions. In those presenting with melena the diagnosis is rarely suspected unless a duplicated segment is found on an ultrasound or barium study. When suspected, a Technetium 99m study helps identify ectopic gastric mucosa within the duplication. However it may not be possible to distinguish from ectopic mucosa in a Meckels diverticulum, another cause of melena.

Short duplications are best treated by resection and primary end to end anastomoses. If complete excision is impossible, long duplications may be separately excised while preserving normal gut. Alternatively the duplication can be partially excised with the remnant stripped of mucosa, but this may be difficult in some cases, as vascularity of the remaining bowel may be compromised.\textsuperscript{1}

This child presented initially with ileal perforation and melena. The perforation was probably secondary to exposure of normal intestinal epithelium to acid secreted by the heterotopic gastric mucosa within the duplicated bowel. As resection was not possible at the time of first operation an elective resection was planned. It could not be carried out as the child was lost to follow up. In retrospect, it might have been better to carry out elective resection of duplicated gut during the first admission itself. This case highlights the emergency presentation of gut duplication and the problems associated with its management.

REFERENCES

Malignant gastrointestinal stromal tumor (neural type) of the rectum

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ABSTRACT
Gastrointestinal stromal tumor is a rare but a new clinical entity. In the gastrointestinal tract it occurs least in the rectum. This tumor is resistant to chemoradiation and adequate surgical resection is the only definitive treatment.

KEY WORDS
Stromal tumor, Malignant, Rectum, Neural type.


INTRODUCTION
Stromal tumors of the gastrointestinal tract are rare neoplasms accounting for <1% of all GI malignancies. They were thought to arise from the muscular layer of hollow organs and their presumed origin from smooth muscle cells has led to the use of terms such as ‘leiomyoma’, ‘leiomyoblastoma’ and ‘epitheloid
leiomyosarcoma’. Unfortunately, the exact origin of these tumors is difficult to determine microscopically. The histogenesis, classification, diagnostic criteria and biological behaviour of these tumors have been subject to much controversy, and the non-specific term gastrointestinal stromal tumor (GIST) has been coined. It has become clear that GISTS show remarkable cellular variability and histology that GIST cells may be able to differentiate towards a variety of cell types or may remain undifferentiated. Immunohistochemistry and electron microscopy have shown GISTS to have myogenic features (smooth muscle GIST), neural attributes (neural GIST), both smooth muscle and neural attributes (mixed GIST) and lack of any differentiation. Very recently, a relationship of GISTS to the interstitial cells of Cajal, a complex cellular network within the muscle wall of the gut where they function as a muscular pacemaker system controlling gut motility, has been proposed. They seem to be resistant to chemoradiation and have a very high potential to recur. Two thirds of GISTS arise from the stomach, 25% from the small bowel (one third in the duodenum), and less than 10% in colorectal regions. Rectal GISTs are extremely rare. This paper reports a case of malignant neural type of a GIST of the rectum, in a young patient.

**CASE REPORT**

A 34-year-old male presented with bleeding per rectum, pain during defecation and evening rise of fever with mild rigor for 6 months’ duration. The pain was dull and constant. He complained of loss of appetite and weight. On examination, the general physical examination was normal. Examination of abdomen was also normal with no hepatosplenomegaly. Proctoscopy revealed second degree hemorrhoids. Digital examination of the rectum revealed a soft swelling touching the examining finger. A clinical diagnosis of carcinoma of the rectum was made. Colonoscopy revealed a large friable growth in the rectum about 5 cm from the anal verge. Biopsy of the lesion suggested undifferentiated adenocarcinoma probably of the signet cell variety. All the investigations, like the blood biochemistry, liver function tests, chest radiography and the abdominal ultrasonography were normal.

Low anterior resection of the rectum with EEA stapling was done. The patient had an uneventful post-operative recovery. The cut specimen of rectum revealed a soft polyoidal growth measuring 5 x 4 x 4 cm projecting into the lumen. The histopathology of the tumor showed sheets of neoplastic spindle-shape cells with oval to elongated vesicular nuclei, prominent nucleoli with eosinophilic cytoplasm. Numerous mitotic figures (12 - 15 / 10 HPF) were seen. The tumor was ulcerated in areas and covered by granulation tissue. The neoplastic cells showed strong positivity with Vimentin and S 100 protein and were negative for Cytokeratin, LCA, Desmin, CD 34 and HMB - 45. The origin of the tumor was from the region of muscularis mucosae. The muscularis propria at the base was not infiltrated. 3 / 7 lymph nodes were positive for tumor and the resected margins were free of tumor.

A final diagnosis of high grade, malignant gastrointestinal stromal tumor of the rectum (neural type) with lymph node metastases was made, based on the tumor marker study and negativity of CD34.

The patient was followed up for over 24 months and he continues to be asymptomatic and healthy.

**DISCUSSION**

Gastrointestinal stromal tumors (GISTS) are the most common mesenchymal tumors of the gastrointestinal tract, but they are rare in occurrence. Majority of patients present in the fifth to the seventh decade of life with male predominance (2:1). They are most common in the stomach (70%), followed by small intestine (20%), colon and rectum (5%) and oesophagus (<5%).

GISTs may range in size from few millimeters to over 30 cm, however, size alone does not predict biologic behavior with certainty. Symptoms usually depend on the tumor size and location but many are asymptomatic, and the tumors are often discovered incidentally during laparotomy for other conditions and they are more benign in variety. Most patients with malignant stromal tumors are symptomatic, the most common being abdominal mass closely followed by GI bleeding as a result of overlying mucosal ulceration and pain. The remainder of the symptoms may include anorexia, dysphagia, obstruction, perforation, or fever. Tumor in the stomach and small intestine commonly present with bleeding. In the oesophagus, the first manifestation may be obstruction, dysphagia and in the rectum, bleeding, obstruction and altered bowel habits. Occasionally, duodenal GISTS may cause obstructive jaundice. GISTS are characteristically well circumscribed and may grow in an endophytic fashion and potentially compromise bowel lumen patency. Malignant GIST tumors occur with increasing frequency in the distal small bowel.

The malignant potential of these tumors are best
estimated by the simultaneous evaluation of several clinical parameters such as size, location in the gut, invasion of the adjacent organ, mucosal invasion, degree of cellularity, cellular architecture, mitotic count, nuclear polymorphism, necrosis and proliferation rate. 

Since within a tumor, there may be considerable heterogeneity with respect to those features that separate benign tumors from malignant ones, thorough sampling for microscopic evaluation is essential, for precise diagnosis. A minimum of one tissue section per centimeter of tumor diameter is required.

At laparotomy, the only absolute criterion of malignancy is the spread beyond the organ of origin at the time of diagnosis, but mucosal ulcer over the tumor is considered a sign of malignancy. Those tumors are locally confined at diagnosis and found incidentally during surgery generally behave in a benign fashion. However, resection of these tumors is necessary, since their behavior is unpredictable and they have to be pathologically examined. Radiation therapy and chemotherapy have been used to a lesser extent, mainly in a palliative setting. Neither modality has been shown to be particularly effective because these tumors seem to be resistant to chemoradiation.

In recurrent tumors, surgery should be reserved largely for symptom control, since disease specific survival seems to be determined by the biology and size of the primary tumor.

T 1571 (Glivec) a potent tyrosine kinase inhibitor which is also a potent inhibitor of C Kit has shown good tolerance and appreciable anti-tumor activity in the GIST. This drug has been used in doses of 400 to 4000 mg daily with good results.

Hurlimann et al observed smooth muscle differentiation in 30% of the cases, neural differentiation is 10%, dual smooth muscle and neural differentiation in 3% and no obvious differentiation in 40%. In tumors with neural differentiation, Vimentin is expressed in 95% of tumors, Neuron specific enolase in 50 - 100 %, Synaptophysin in 100%, Neurofilament protein in 10%, S-100 in 20 - 60%, Vasointestinal peptide in 20 - 40% and CD 34 in 60%. Tumors with smooth muscle differentiation, do not express Neuron specific enolase, Synaptophysin, Chromogranin, Glial fibrillary acidic protein, or Protein gene product 9.5 but express lineage specific markers, like Muscle specific antigen (HHF - 35) in 68%, Smooth muscle actin (SMA) in 57% and Desmin upto 50%.

A type of GIST showing neural differentiation is the gastrointestinal autonomic nerve tumor (GANT), which are uncommon stromal tumors with morphological features resembling the cell processes of the enteric autonomic plexus that occasionally develops in the context of von Recklinghausen’s disease. GANTs are typically epithelioid or spindle celled and usually of low histological grade. They typically express S-100, Neuron specific enolase, Vimentin and Synaptophysin and they are CD 34 negative, as in the present case. GANTs can be distinguished from other GISTs only on the basis of their unique ultrastructural features.

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Kaposi’s sarcoma in a follow-up patient of malignant schwannoma after seroconversion

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