



Case Report

Gastrointestinal Stromal Tumor Of Rectum

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ABSTRACT

This is a case report of a 40 year old lady presenting with gastrointestinal stromal tumor of rectum. Gastrointestinal stromal tumors (GIST) tend to present with higher frequency in the stomach (60-70%) and small bowel (20-25%) while colon and rectum represent rare sites. Gastrointestinal stromal tumors of the rectum represent around 5% of all GISTs and 0.1% of all tumours originating in the rectum. Anterior resection of rectum with stapled anastomosis with protective colostomy was done in our patient and Imatinib [400 mg per day] was advised as adjuvant therapy. To date, follow up of the patient has been satisfactory. The diagnostic work up of rectal GIST is similar to other rectal tumors. Complete surgical resection is the principal curative treatment for primary rectal GIST. The efficacy of adjuvant imatinib therapy needs further evaluation.

KEYWORDS: Gastrointestinal stromal tumor, Rectal GIST

INTRODUCTION

Gastrointestinal stromal tumour (GIST) is a rare tumour involving the gastrointestinal tract and expresses CD117, a c-kit proto-oncogene, which can be detected immunohistochemically.[1,2] The most common sites for GIST are the stomach (60%-70%) followed by the small intestine (20%-25%), whereas only about 5% of all GISTs occur in the rectum. Rectal GISTs make up 0.1% of all tumours originating in the rectum. The symptoms of rectal GIST do not differ from those of other rectal tumours and diagnostic work up is also similar to that for any other rectal mass. Because of the lower incidence of rectal GIST, the clinicopathological profiles have not yet been

accurately characterised, and there is therefore the tendency to validate the same prognostic factors as for GIST at other sites, particularly gastric GIST.[3,4,5] Complete surgical resection with negative tumour margins is the principal curative procedure for primary rectal GIST, particularly for those at a low risk.[6,7,8] Imatinib therapy has long been regarded as a treatment for metastatic GISTs. Its role as adjuvant therapy for primary resectable rectal GIST is controversial. But recent studies have advised routine use of imatinib as an adjuvant treatment after complete resection of primary high grade rectal GIST.[9,10] In this report, the authors describe a rare case of primary rectal GIST.

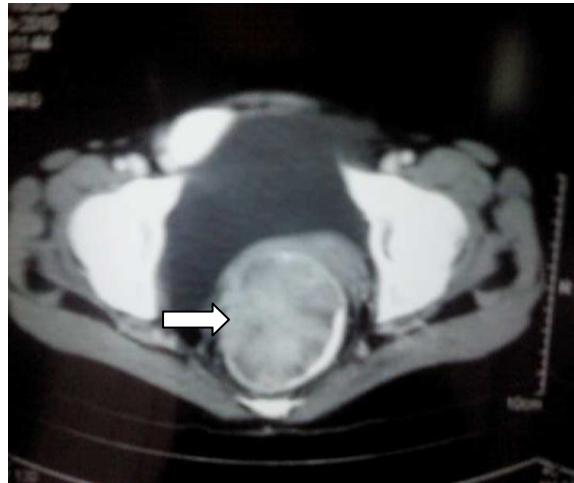
CASE REPORT

A 40 year old female presented to surgical oncology department at our institute with bleeding per rectum and painful defecation for 3 months. Clinical examination did not detect any palpable abdominal mass. There was no palpable rectal mass on digital rectal examination.

Basic investigations were within normal limits. Serum CEA and CA19.9 values were normal.

Chest Xray was normal. On colonoscopy, luminal polypoidal growth was present 10cms from anal verge. Scope was not negotiable beyond the growth. Multiple biopsies were taken . Biopsy showed malignant spindle cell neoplasm of rectum. CT scan of abdomen & pelvis showed polypoidal growth projecting into the lumen from anterior wall of rectum. There was no evidence of pelvic lymphadenopathy. Liver was normal.

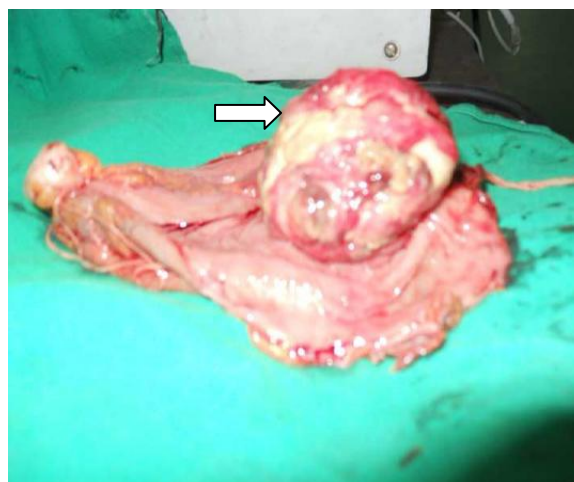
Figure 1: CT scan showing polypoidal growth from anterior wall of rectum



An explorative laparotomy was done which showed a 7 × 6 cm intraluminal tumor in upper rectum. No enlarged nodes were found in pelvis. Liver was normal. There was no evidence of free fluid. Anterior resection of rectum with stapled anastomosis with protective colostomy was done. The postoperative period was normal and patient

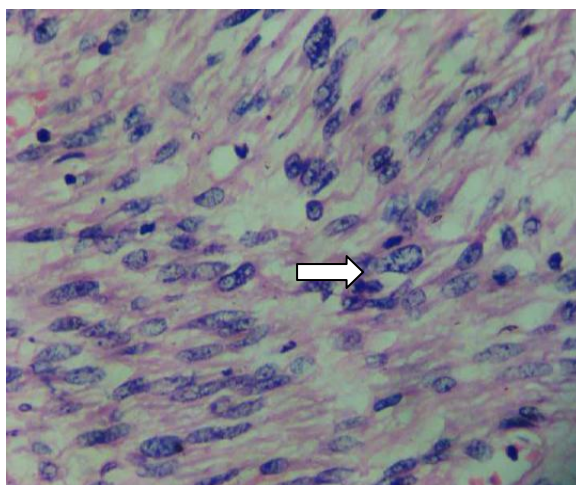
was sent home on eighth day. The patient was advised adjuvant therapy with imatinib [400 mg per day]. The colostomy was closed two months after surgery. On gross sectioning a 7 × 5 × 5 cm polypoidal lesion was seen arising from mucosa of the rectum . Cut margins were away from the tumor.

Figure 2: Gross appearance of resected anterior rectal growth



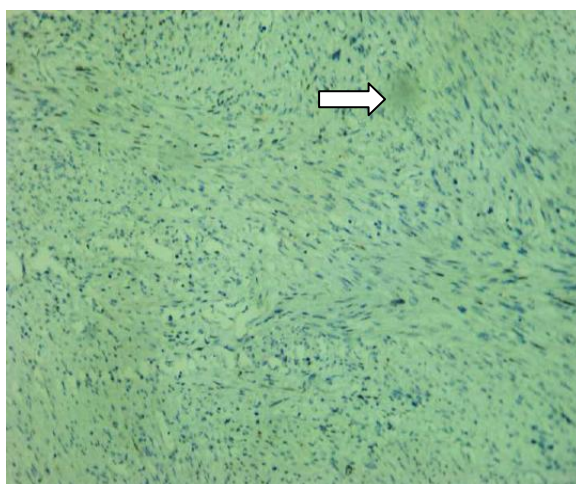
Histopathology showed high grade gastrointestinal stromal tumor of rectum with distal & proximal cut margins being free of tumor.

Figure 3: High power view showing spindle cells



Diagnosis was confirmed by the immunohistochemistry. CD117, CD34 and SMA were strongly positive and the mitotic index was $> 50/50$ per hpf.

Figure 4: Immunohistochemistry - CD 117 strongly positive



After 11 months of regular follow up, she showed no signs of either local recurrence or distant metastasis.

DISCUSSION

GIST is a mesenchymal tumour of gastrointestinal tract thought to arise from interstitial cells of Cajal that are normally part of the autonomic nervous system and expresses CD117, a tyrosine kinase growth factor receptor.[1] Most GISTs originate within the muscularis propria and most commonly have an exophytic growth pattern. The symptoms of rectal GIST do not differ from those of other rectal tumours and diagnostic work up is also

similar to that for any other rectal mass. A focal, well circumscribed mural mass is the most common finding on CT scan or MRI of the pelvis. Furthermore, CT or MRI scanning is needed to detect local invasion and presence of possible metastasis.

Preoperative biopsy plays a key role in the diagnosis of GIST, since it provides information on the immunohistochemical features and mitotic

count. GIST typically expresses CD117, often CD34 and sometimes SMA and S-100, but their expression vary depending on different sites. Miettinen et al[3] found that CD34 expression in rectal GIST is 92%, but only 50% in small intestinal GIST. Smooth muscle antigen (SMA) is most frequently seen in small intestinal GIST (47%), whereas it has been observed in only 14% of rectal GISTs. The reason for these variations has not yet been explained.

Because of the lower incidence of rectal GIST, the clinicopathological profiles have not yet been accurately characterised, and there is therefore the tendency to validate the same prognostic factors as for GIST at other sites, particularly gastric GIST. The most important and easily applicable histological criteria for prognosis of GIST are its size and mitotic rate.[4] A rate of ≤ 5 mitoses per 50 hpf is commonly used as a limit for a tumour with expected benign behaviour or low risk of malignancy.[4] Tumours of < 2 cms in diameter are generally expected to behave in a benign fashion. Tumours of < 5 cms in diameter are associated with a better survival rate than those of 5-10 cms in diameter, which in turn have a better prognosis than those of > 10 cms in diameter.[4] Degrees of cellularity and atypia have also been suggested as useful criteria, but their reproducibility is more problematic. The epithelioid phenotype, which seems to lead to a worse outcome, together with symptoms lasting for at least a year, might be considered as other prognostic factors.[4]

Complete surgical resection with negative tumour margins is the principal curative procedure for primary rectal GIST, particularly for those at a low risk. Various surgical procedures may be considered including local excision, anterior resection of the rectum and abdomino-perineal resection. The choice of procedure depends on tumour size and location. [6,7,8]

Imatinib therapy has long been regarded as a treatment for metastatic GISTs. Its role as adjuvant therapy for primary resectable rectal GIST is controversial. But recent studies have advised routine use of imatinib as an adjuvant treatment after complete resection of primary high grade rectal GIST.[9,10]

CONCLUSION

Rectal GIST should be included in differential diagnosis of rectal tumors although it is rare. The diagnostic work up of rectal GIST is similar to other rectal tumors. Immunohistochemistry is an essential part of diagnosis. Complete surgical resection is the principal curative treatment for primary rectal GIST. The efficacy of adjuvant imatinib therapy needs further evaluation.

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