

Malignant gastrointestinal stromal tumor of the rectum with prolapse

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Gastrointestinal stromal tumors are rare tumors of the gastrointestinal tract, and they account for less than 1% of gastrointestinal neoplasms. Most of these tumors may not originate from smooth muscles. With the advent of electron microscopy, interstitial cells of Cajal, a pleuropotential intestinal pacemaker cell, was found as the originator of gastrointestinal stromal tumors.¹ The stomach is a common site for gastrointestinal stromal tumors, occurring in 50-60% of cases, followed by the small intestine, accounting for 25% of cases. Less than 10% of these tumors involve the colon and rectum.² Prolapse of the tumor is a rare presentation in cases of rectal stromal tumors. A few cases have been reported in the literature. Herein, we are presenting an interesting case of a malignant stromal tumor of the rectum with prolapse.

A 36-year-old gentleman presented to the emergency room with complaints of protrusion of a mass out of the anal verge, associated with bleeding per rectum for a few hours. He was seen in another hospital with the same complaint, one year earlier. At that time, the prolapse was reduced and biopsy from the mass showed stromal leiomyosarcoma. He was advised surgery, which he refused. Since then he has had 4 episodes of prolapse, which were reduced every time. On examination he was in pain. His vital signs were stable. He looked pale. The abdomen was soft. Local examination showed a large mass, of 10 cm, protruding out of the anal verge. It was nodular, ulcerated on the medial surface, and bleeding to touch (Figure 1). His hematological investigations showed, hemoglobin 6.7 g/dl, white blood cell count 12000/UL, and platelets 355000/UL. His liver function test, and coagulation profile were within normal limits. He was transfused with 4 units of packed red blood cells. The prolapse of the mass was reduced under general anesthesia, and biopsy was taken. Histopathology showed a malignant gastrointestinal stromal tumor of the rectum. A CT scan of the abdomen and pelvis showed a malignant rectal mass, with multiple liver metastasis. On exploration he was found to have multiple liver metastasis, omental metastasis, and a large mass in the rectum. A palliative abdomino perineal resection with a colostomy was carried out, as the tumor reached the anal canal, and it was not possible to save the sphincter. His postoperative course was unremarkable, and he was discharged after 2 weeks, with a follow up appointment in the surgical and oncological clinics. Histopathology of the specimen showed a malignant



Figure 1 - Stromal tumor of rectum with prolapse.

gastrointestinal stromal tumor of the rectum, with negative proximal, and distal resection margins. Omental biopsy showed metastasis to the omentum.

Gastrointestinal stromal tumors of the rectum are rare tumors, accounting for less than 5% of gastrointestinal stromal tumors. Tumors involving the rectum commonly present as bleeding per rectum, constipation, and a rectal mass, but rarely with rectal tumor prolapse.³ Helpful investigations for pre operative diagnosis includes, lower gastrointestinal endoscopy, CT scan, MRI, and endoscopic ultrasound. Endoscopic ultrasound has a sensitivity of 80-100% in detecting these tumors. Endoscopic ultrasound findings can be used to differentiate benign from malignant stromal tumors. Features suggesting malignancy include size more than 4 cm, irregular margin, echogenic foci, and cystic spaces.¹ Radiological investigations are important, as they detect the metastasis, and delineate the tumor, which may allow a sphincter preserving procedure. Electron microscopy and immunohistochemical techniques are required for the final diagnosis of these tumors. Although the colon and rectum are less common sites, the majority of the gastrointestinal stromal tumors of the colon and rectum are malignant, and likely to develop metastasis. Therefore, these tumors are associated with poor prognosis.⁴

Surgical treatment is the treatment of choice for gastrointestinal stromal tumors. Abdominoperineal resection has been the treatment of choice for tumors of the rectum. Recently, transanal excision, or local excision of small confined tumors has been advocated to preserve continence. The aim of surgery should be complete resection with negative resection margins, without lymph adenectomy.⁴ The recent discovery of expression of C-kit receptor tyrosine kinase in these tumors, has led to the tyrosine kinase inhibitor Glivec, as molecular targeted therapy for these tumors. Glivec is indicated in patients with unresectable tumors, or with metastatic disease.⁵

Clinical Notes

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References

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