Atypical Presentation of a Stromal Tumor of the Small Gut

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Citation

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Abstract

An atypical presentation of a stromal tumor of the small gut with infiltration of the urinary bladder and formation of a pseudo-bladder in a 58-year-old female patient is described. Operative treatment included resection and construction of an ileal conduit. Clinical management of this disease and recent reports in the literature are discussed.

CASE REPORT

A 58-year-old lady was admitted in our hospital in August 2005 with complaints of fecalurea, weight loss, and generalized weakness for more than 1 year duration. The patient was admitted for evaluation and put on intra-venous fluids, bowl rest, three way catheter with continuous saline irrigation, antibiotics, multivitamins and amino acids. A baseline assessment was made with a provisional diagnosis of colovesical fistula possibly secondary to diverticulitis, tuberculosis or colon carcinoma. Diverticulitis was excluded because of the rarity of the disease in our region. Tubercular profile, colonoscopy, and ultrasonography all were normal and did not give a significant clue to diagnosis. A CECT of the abdomen did only provide indirect evidence of small bowl involvement but again no preoperative diagnosis could be made. In spite of being on continuous saline irrigation through a three way catheter for many days the patient was still always forming feculent urine.

She was subjected to exploratory laparotomy which revealed a mass in the pelvis formed by loops of ileum, by sigmoid colon and partly by the uterus. Once the loops were teased, they broke as flimsy adhesions, revealing the tip of the urinary catheter. On examination in detail, we found that the urinary bladder was completely eaten up by a diffuse ulcerative growth arising from small gut, only the trigone was spared. Two loops of ileum anteriorly, colon on the side and uterus posteriorly had almost formed a pseudo-bladder by flimsy adhesions with each other and around the trigone. Small gut loops were continuously pouring all the contents into this cavity while the sigmoid colon had a doubtful communication with the said cavity. A segmental resection of small gut segments and the sigmoid colon area was planned, and the entire diseased segment of the gut was removed and anastamosed end to end. The case was discussed with an urologist to plan a urinary diversion and with his help an ileal conduit with implantation of both ureters on a stent was made. With the help of a gynaecologist a hysterectomy was performed and the specimen was sent for histopathological examination. Immunohistochemistry (CD 117, c-kit reactive (positive)) confirmed the morphologic diagnosis of a malignant stromal cell tumor.

The patient was discharged on the tenth postoperative day but unluckily developed serious depression on follow-up and refused further visits at the department of medical oncology. The ileal conduit was working nicely. The patient died after 9 months secondary to depression and probably to the disease process.

Figure 1

Figures 1-3: Stromal tumor of the small gut. Resection and construction of an ileal conduit. Intraoperative views.

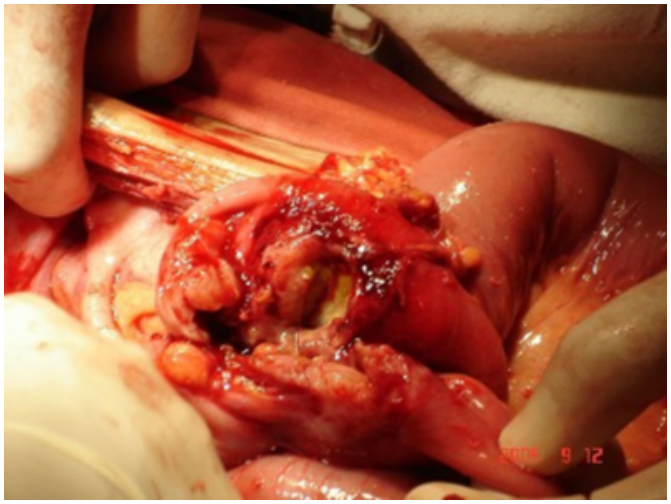


Figure 2

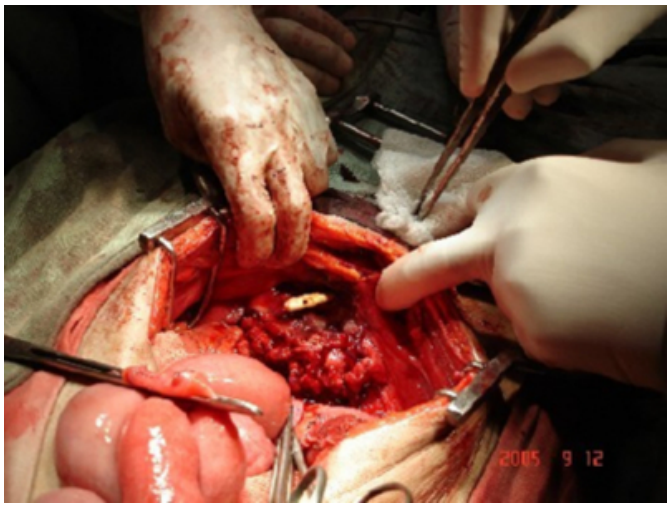
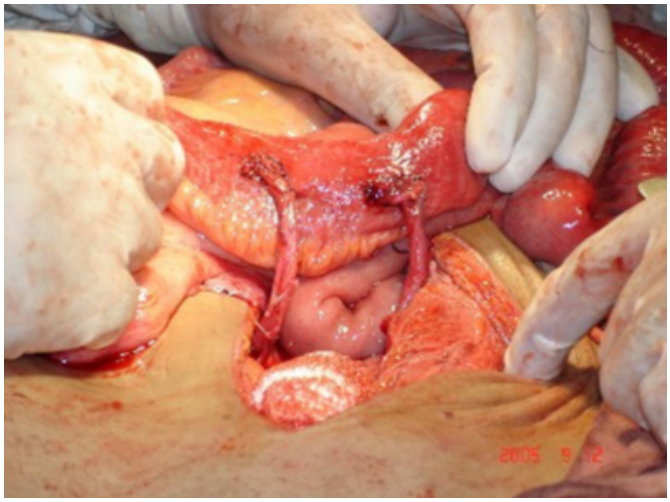


Figure 3



DISCUSSION

Malignant gastro-intestinal stromal tumors (M-GISTs) are rare mesenchynal tumors originating in the wall of the gastrointestinal (GI) tract. Most patients with M-GIST of the small intestine relapse following resection but survival may be prolonged. In univariable analysis, stage at presentation and complete resection were significant prognostic variables for overall survival; grade was not significant. Localized and locally advanced M-GISTs of the small intestine have a mean overall survival of more than 5 years. Complete resection should be the goal of initial surgical treatment.[1] The actual cell of origin of GIST is a pluripotent mesenchymal stem cell programmed to differentiate into the interstitial cell of Cajal which is a GI pacemaker cell; hence, these tumors are also called GI pacemaker cell tumors[2].

Using the histologic classification of Fletcher[3] et al., the tumors are divided into 2 groups. Tumors classified as very low, low and intermediate risk comprise one group. The second group consists of tumors classified as high risk. The 5-year survival rates were 63% in the former group and 34% in the latter group[3]. Gain-of-function mutations in exon 11 of the C-kit protoncogene are associated with most GISTs. These mutations lead to constitutive over-expression and autophosphorylation of C-kit, provoking a cascade of intracellular signaling that propels cells towards proliferation or away from apoptotic pathways. This discovery was a landmark elucidation of the etiology of a disease on a molecular level[4]. GISTs are also a feature of the rare Carney triad which consists of epitheloid gastric stromal tumors, pulmonary chondromas, and extra-adrenal paragangliomas[5]. Usually the information gathered from investigations to diagnose GIST may be limited, however, specialized

investigations like contrast CT may help in diagnosing and staging. Ghanem[6] et al. performed CT scanning on patients with histologically confirmed primary (n = 20) or recurrent (n = 16) GISTs. They described the CT characteristics of GISTs; dividing them into small (<5cm), intermediate (5-10cm), and large (7-10cm) tumors. Small GISTs were sharply demarcated, homogeneous masses, mainly exhibiting intraluminal growth patterns. Intermediate types were characterized by irregular shape, heterogeneous density, an intraluminal and extraluminal growth pattern and signs of biological aggression, including adjacent organ infiltration in 9 primary and 2 recurrent lesions. Large GISTs featured irregular margins, heterogeneous densitities, locally aggressive behavior and distant and peritoneal metastases[6]. In 1997, Shojan and colleagues described a GIST appearing hypointense on T_2-weighted images[7]. Position emission tomography scanning has recently been termed as an excellent study for detecting metastatic disease. It has also been used to monitor responses to adjuvant therapies such as imatinib mesylate[8].

CONCLUSION

This is a very unusual and a very rare type of presentation of M-GIST; probably the first of its kind reported in literature where the urinary bladder was completely eaten up and replaced by a pseudo-bladder.The patient had reported quite late because of social reasons. The patient felt ashamed to report her problem of fecalurea even to her doctor son. We also want to advise doctors and patients that complaints of fecalurea can be sometimes secondary to a devastating malignancy of small gut which - if left untreated - can prove disastrous for the patient.

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