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Small bowel sarcoma: A rare case report

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CASE STUDY

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ABSTRACT

Sarcoma of the small bowel is an extremely rare type of small bowel malignancy and represents around 10 per cent of small bowel cancers. Usually asymptomatic, patients can present with chronic abdominal distention with pain and/or weight loss. Most pathologists now refer to small bowel sarcomas (SBS) as gastrointestinal stromal tumours as they are mesenchymal neoplasms believed to be derived from the interstitial cells of Cajal in the gastrointestinal tract. SBS can be highly vascular and commonly ulcerate and/or bleed. They tend to be aggressive and have a poor prognosis. Surgery is the treatment of choice for SBS, with definitive diagnosis usually made postoperatively with the aid of histopathology. We would like to highlight the importance of careful intra-operative examination of an unknown small bowel (SB) mass, which can give the surgeon clues to the type of tumour present. This case report aims to demonstrate this important process: recognising SBS intra-operatively will aid the surgeon with performing the appropriate resection as minute omental metastasis can be present with SBS; in such cases an omentectomy during the laparotomy is recommended. In institutions where available, intra-operative radiotherapy is ideal.

Key Words
Small bowel sarcoma, small bowel malignancy, small bowel tumour

Implications for Practice:

1. What is known about this subject?
Small bowel sarcomas (SBS) represent around 10 per cent of small bowel cancers.

2. What new information is offered in this case study?
Careful examination of an unknown small bowel (SB) mass can help the surgeon identify the type of tumour present. This case report highlights this identification process.

3. What are the implications for research, policy, or practice?
Resection with adequate margins is required with omentectomy to reduce the chance of recurrence. Great care must be taken to prevent tumour spillage as this has been shown to be a mode of metastasis.

Background

Sarcoma of the small bowel (SB) is an extremely rare type of SB malignancy and represents around 10 per cent of SB cancers.[1] Adenocarcinomas are the most common followed by carcinoid tumours, sarcomas, and lymphomas.[2] Small bowel sarcomas (SBS) arise from transformed cells of mesenchymal origin. In the early stages, they can be asymptomatic with patients usually presenting with chronic abdominal distention with pain and/or weight loss. A palpable abdominal mass is present in 66 per cent of SBS.[3] SBS are mostly leiomyosarcomas and tend to be large extramural masses, often with a myxoid cut surface.[3] Most pathologists now refer to SBS as gastrointestinal stromal tumours as they are mesenchymal neoplasms believed to be derived from the interstitial cells of Cajal in the gastrointestinal tract. SBS can be highly vascular and commonly ulcerate and/or bleed. They tend to be aggressive and have a poor prognosis. This case report aims to educate the medical community on this rare condition and highlight subtle clues that can help the surgeon identify SBS intra-operatively.

Case details

A 69-year-old woman presented to our emergency department with a syncopal episode associated with acute onset generalised abdominal pain and gradual distention of

her abdomen. She reported symptoms associated with small bowel obstruction ongoing for the past 24 hours. She had a background history of chronic anaemia and was taking oral iron replacement therapy. A plain abdominal X-ray demonstrated dilated small bowel loops and a CT scan was subsequently performed. The CT showed thickened loops of small bowel with an aneurysmal dilation. This was in continuation with a large cystic mass in the mid-jejunum with free gas under the diaphragm (Figures 1 and 2).

Figure 1: CT scan of the large cystic mass in continuation with the small bowel

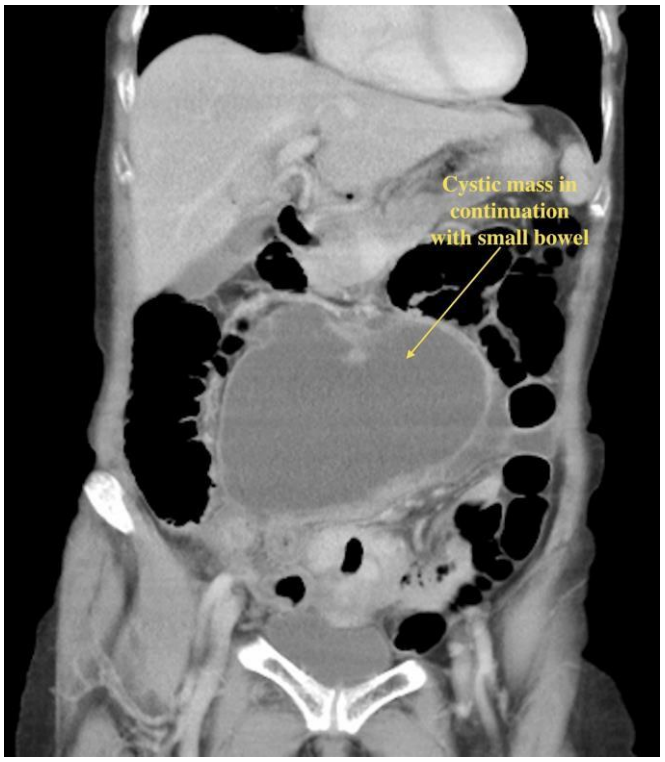
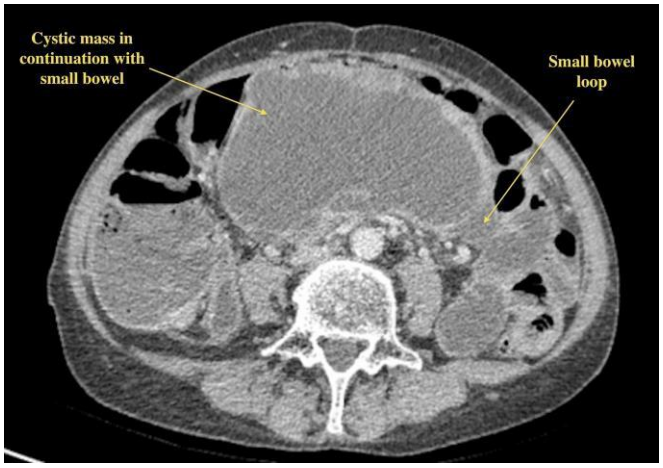


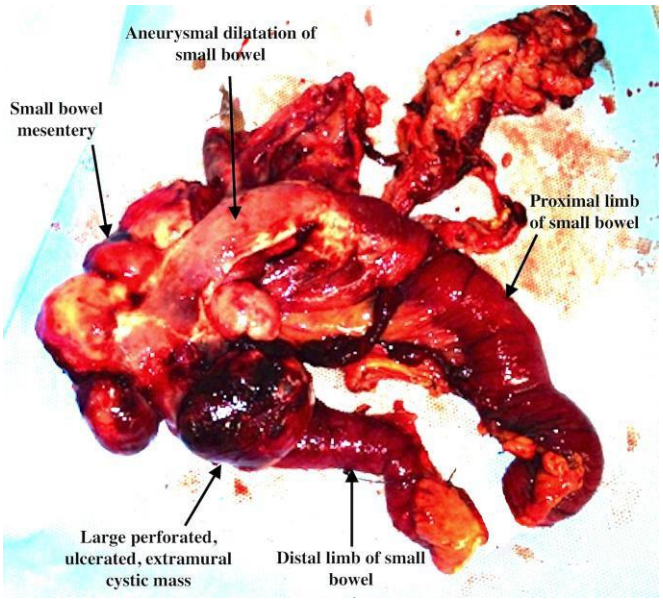
Figure 2: CT scan of the SBS in association with the small bowel



Differential diagnosis at this point was of either complicated inflammatory bowel disease, an abscess associated with localised perforation, or a small bowel lymphoma. Intra-operatively we found a large amount of blood and clots within the peritoneal cavity with significant ongoing bleeding from a necrotic, perforated mass in the mid-jejunal wall. This mass was resected with adequate surgical margins. The size of the entire specimen was 380mm in length and 28mm in maximal diameter. On gross examination of the tumour, it appeared to be a firm whitish/tan lobulated mass firmly adherent to the intestinal wall.

Histology showed a tumour 110mm in size proving to be a high-grade sarcoma with smooth muscle differentiation and tumour necrosis arising within the intestinal wall with extension into the overlying mucosa. Immunohistochemistry performed showed the tumour cells were positive for smooth muscle Actin, S100, and MITF-1. Melan A, HMB45, CD117, CD34, CD31, ALK1, Desmin, and cytokeratins were negative. The patient's postoperative recovery was uneventful. She was referred to the oncology unit for further management and radiotherapy. The resected specimen is shown in Figure 3. This section of the mid-jejunum shows the whitish/tan tumour 150mm from the distal margin and 195mm from the proximal margin.

Figure 3: Resected specimen of the mid-jejunum, with the 110mm whitish/tan extramural cystic mass



Discussion

SBS can be very difficult to histologically classify by grade and stage.[1] Insufficient patient data has hindered the accurate determination of survival factors of patients with SBS. Although SBS are slow to metastasise, approximately 30 per cent of patients were found to have metastasis at the time of laparotomy.[4] This occurs either by direct

peritoneal implantation of tumour cells, blood-borne metastases, and less frequently by lymphatic dissemination. Surgery is the treatment of choice for SBS, with definitive diagnosis usually made postoperatively with the aid of histopathology. O'Rourke et al.[3] outlined the importance of intra-operative identification of the type of small bowel neoplasm based on its macroscopic appearance. Careful intra-operative examination of the mass can give the surgeon clues about the type of SB mass. Minute omental metastasis can be present, hence an omentectomy during the laparotomy is also recommended.[4] Their descriptions aid the surgeon intra-operatively in regard to the patient's further management.

O'Rourke et al. also highlighted that SBS commonly occur in the jejunum, are usually greater than 5cm, and appear to be a solitary extramural fleshy mass and whitish/purple in colour. Slow and methodical examination of the specimen is important. Adenocarcinomas, lymphomas, and carcinoid lesions in contrast, tend to be mucosal, transmural, and submucosal, respectively. Being able to examine and diagnose an unknown small bowel mass is a useful tool. Although the patient recovered well and received appropriate medical treatment, we may have performed a partial omentectomy if we had suspected that this unknown mass was a sarcoma.

Small bowel sarcomas respond well to radiotherapy and usually require a higher than normal dose.[5] Intra-operative radiotherapy, when available, is an excellent option as it provides the major advantage of being able to administer high doses of radiation to a localised area without exposing other unaffected organs to the radiation.[5]

SBS are slow growing but aggressive malignancies. Patients often present with an acute perforation from the ulcerated tumour and or intra-abdominal bleeding. Approximately 30 per cent of patient will have localised metastasis at the time of laparotomy. Investigations to stage SBS are advisable either prior to or immediately post-laparotomy as this may impact the patient's management. Careful examination of an unknown SB mass can give the surgeon clues about the type of tumour present. Macroscopically, SBS tend to be extramural in appearance. Resection with adequate margins is required along with an omentectomy to reduce the chance of recurrence. Great care must be taken to prevent tumour spillage as this has been shown to be a mode of metastasis. Intra-operative or post-operative radiotherapy has shown to be of great benefit.

Conclusion

Small bowel sarcomas are slow growing but aggressive malignancies. Intra-operative examination of the small bowel mass can help the surgeon identify the type of tumour. An omentectomy during the laparotomy is also recommended. Small bowel sarcomas respond well to radiotherapy and should be used when available. Representing 10 per cent of all small bowel cancers, surgeons should be aware of SBS be able to identify this condition intra-operatively.

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PEER REVIEW

Not commissioned. Externally peer reviewed.

CONFLICTS OF INTEREST

The authors declare that they have no competing interests.

PATIENT CONSENT

The authors, *Durgakeri P, Sarkar A, and Jones B,* declare that:

1. They have obtained written, informed consent for the publication of the details relating to the patient(s) in this report.
2. All possible steps have been taken to safeguard the identity of the patient(s).
3. This submission is compliant with the requirements of local research ethics committees.