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Case Report

A fortunate presentation of intestinal obstruction secondary to a rare sarcomatoid tumour of the small bowel

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ABSTRACT

Intussusception in the adult is rarely from a benign cause and is almost always pathological. Causes include carcinomas, polyps, Meckel's diverticulum, or colonic diverticulum. Common symptoms include abdominal pain, intestinal obstruction, palpable abdominal mass, gastrointestinal (GI) bleeding, and anaemia. Sarcomatoid carcinoma is a rare type of small intestinal malignancy exhibiting carcinomatous and sarcomatous features. It primarily affects older patients, mean age 57, and is 1.5 times more prevalent in men. This is an interesting case report of a patient presenting with intussusception secondary to a sarcomatoid tumour of the small bowel. Surgery is the treatment of choice in adults with intussusception due to the high malignancy potential. Furthermore, surgical resection of the affected bowel is the definitive form of therapy as small bowel sarcomatoid tumours are not responsive to chemotherapy and radiotherapy. Early surgical intervention helps reduce mortality as it allows for early staging, treatment, and monitoring of the tumour. The patient was fortunate to have presented with intussusception, facilitating early surgical intervention, and was found to have a low disease stage.

Keywords: General surgery, Small bowel tumour, Imaging, Unique

INTRODUCTION

Intussusception in the adult is rarely from a benign cause and is almost always pathological. Causes include carcinomas, polyps, Meckel's diverticulum, or colonic diverticulum. Common symptoms include abdominal pain, intestinal obstruction, palpable abdominal mass, GI bleeding, and anaemia. Sarcomatoid carcinoma is a rare type of small intestinal malignancy exhibiting carcinomatous and sarcomatous features. It primarily affects older patients, mean age 57, and is 1.5 times more prevalent in men.[1]

This is an interesting case report of a patient presenting with intussusception secondary to a sarcomatoid tumour of the small bowel.

CASE REPORT

A 32-year-old female presented to an emergency department with two weeks of worsening intermittent right upper quadrant (RUQ) pain, exacerbated by eating and ibuprofen. It improved temporarily with an oral PPI. She was systemically well, and her examination was unremarkable. Her blood tests showed a C-reactive protein (CRP) of 30 and were otherwise unremarkable, including beta-human chorionic gonadotrophin (BHCG) of <1.2. Her medical history included obesity and well controlled asthma. She worked as a registered nurse and had no lifestyle risk factors. She previously had an abdominal ultrasound for similar pain as an outpatient which was negative, and this was repeated only showing mild CBD prominence. She was subsequently discharged with ongoing PPI therapy for further GP review.

She represented 4 days later with ongoing pain and now with bilious post prandial vomiting and not passing wind. A computed tomography (CT) abdomen demonstrated a closed loop jejunal volvulus (Figure 1). She was reviewed by the surgeons and underwent a diagnostic laparoscopy finding an intussusception 10 cm from duodenojenunal flexure. This could not be reduced and the operation was converted to a laparotomy + jejunal resection. A hard area was noted within the intussusception and a large growth as lead point was identified on review of resected specimen.

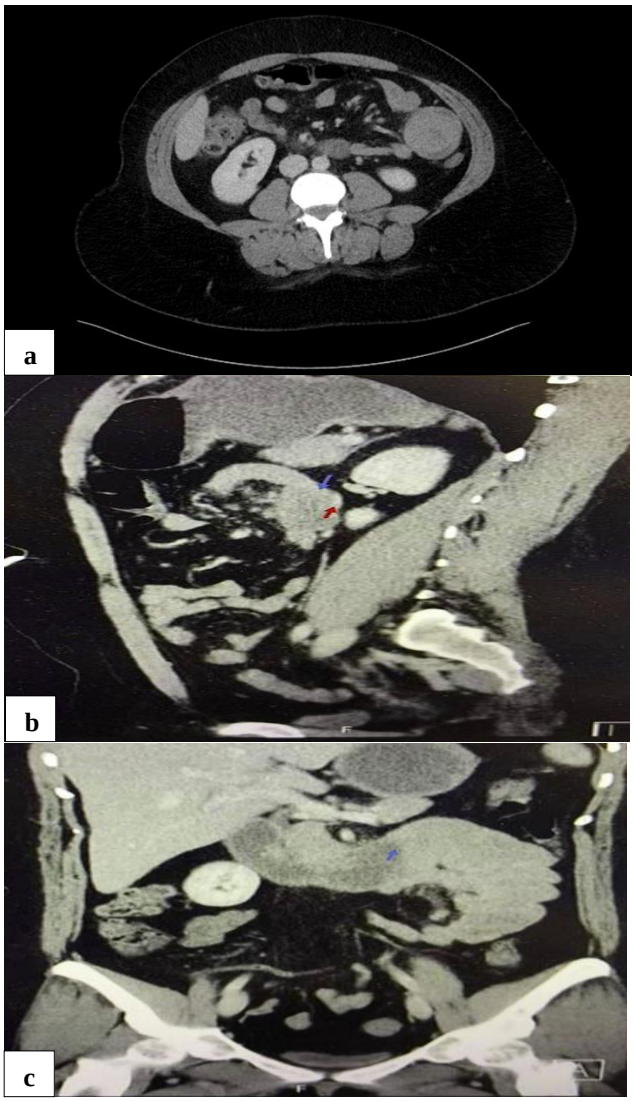

Figure 1: (a) CT abdomen demonstrating intussusception of the proximal jejunum with no evidence of a mass (red arrow); (b) CT abdomen on a sagittal view demonstrating intussusception of the proximal jejunum. The blue arrow demonstrates the intussuseptum while the red arrow represents the intussucipiens; and (c) CT abdomen on a coronal view demonstrating obstruction at the proximal jejunum (blue arrow).

Macroscopically, a large polypoid tumour 40×25 mm with the tumour well clear of mucosal margins. Microscopy showed a spindle cell neoplasm arising from the submucosa, intact serosa and variable morphological features. Immunohistochemistry showed positive stains to CK, CD34 and SMA. Features were consistent with high grade sarcomatoid neoplasm.

On day 5 post operation she was discharged after methylene blue staining confirmed no leak from the functional end to end anastomosis. Further staging with a chest CT showed no evidence of intrathoracic metastatic disease. She was referred to oncology for further follow-up and had no evidence of metastasis during the one year follow up.

DISCUSSION

Intussusception in the adult is rarely from the benign cause and is almost always pathological. Causes include carcinomas, polyps, Meckel's diverticulum, or colonic diverticulum. From this, 65% of cases are secondary to malignancy. Sarcomatoid carcinoma, initially described by Dikman and Toke, is a rare type of small intestinal malignancy exhibiting carcinomatous and sarcomatous features.[1] To date, only 32 cases have been reported in the literature, with our case being unique in three ways; our patient is the youngest in the series, this is the first reported Australian case of a small bowel sarcomatoid tumour, and this is the first report of intussusception secondary to a primary sarcomatoid neoplasm.[2] It primarily affects older patients, mean age 57, and is 1.5 times more prevalent in men.[3] Common sites of involvement within the GI tract are the jejunum, ileum, and duodenum. Macroscopically, the mass tends to be polypoid or fungating/ulcerating with necrotic or haemorrhagic features and has an average size of 7 cm.[2] Common symptoms include abdominal pain, intestinal obstruction, palpable abdominal mass, GI bleeding, and anaemia.[2] In our case, the patient presented with intestinal obstruction.

Surgery is the treatment of choice in adults with intersuception due to the high malignancy potential. Furthermore, surgical resection of the affected bowel is the definitive form of therapy as small bowel sarcomatid tumours are not responsive to chemotherapy and radiotherapy.[2,4] Furthermore, patients with sarcomatoid tumours often present in the late stages of disease (when distant metastasis have developed). As such, 70% of patients die within 2 months to 3 years from diagnosis.[5] Therefore, early surgical intervention helps reduce mortality as it allows for early staging, treatment and monitoring of the tumour. In this case, our patient was fortunate to have presented with intussusception, facilitating early surgical intervention and was found to have a low disease stage.

CONCLUSION

This is the first reported case of a sarcamatoid tumour causing intussusception of the small bowel. Furthermore, she is the youngest patient in literature to have this condition and remains to be well after three years of follow up.

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