



Ileal volvulus and its association with carcinoid tumours

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CASE REPORT

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Abstract

Small bowel volvulus is a rare cause of bowel obstruction in the Western World. It is often divided into primary and secondary causes. This report presents a case of secondary ileal volvulus with underlying carcinoid tumour.

Key Words

Small bowel volvulus, adult, carcinoid

Implications for practice

- 1. What is known about such cases?** Small bowel volvulus is rare and can be divided into primary and secondary causes.
 - 2. What is the key finding reported in this case report?** Laboratory tests and imaging report false negative findings in the presence of small bowel gangrene.
 - 3. What are the implications for future practice?** All adult patients who present with small bowel volvulus should have malignancy and carcinoid tumours considered as a cause.
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Background

Volvulus occurs when a segment of bowel rotates around itself or its mesentery causing a partial or closed obstruction of its lumen. Small bowel volvulus is rare and only few cases have been reported worldwide. Small bowel volvulus accounts for approximately 3-6% of all cases of small bowel obstruction in the western world.^{1,2} This report presents a case of ileal volvulus associated with carcinoid tumour, its presentation, investigation findings and a discussion of its management.

Case details

A 70-year-old elderly lady presented to the emergency department with an acute onset of epigastric pain, which came in waves radiating to bilateral hypochondria. The pain was associated with nausea and dry retching. She was diaphoretic and reported feeling light-headed. No other associated symptoms were reported. Her last bowel motion was earlier in the morning. She was previously well. Relevant past medical history included sleep apnoea, which was managed with the aid of a CPAP machine at night. There was no previous surgical history. Family history included ischemic heart disease and hypertension.

Laboratory test results on admission were as follows; Hb 123 g/L, WCC 9.9 x 10⁹ with neutrophils of 6.99 x 10⁹. Normal renal function. ALT 52 U/L, ALP 85 U/L, GGT 54 umol/L, bilirubin 8 g/L. Troponin < 0.04 ug/L, lipase 111 U/L venous lactate of 1.1, glucose 8.7. Her chest X-ray was reported as normal and a CT angiogram of her chest and abdomen was reported as finding of small perihepatic fluid collection. These tests were non-specific for her life threatening condition. She was admitted to the High Dependency Unit for further observation under a medical team with surgical input. The working differentials were possible atypical presentation of myocardial infarction, small bowel obstruction or pancreatitis. She continued to deteriorate and on review eight hours after presentation, generalised peritonism had ensued with a palpable mass in the left upper quadrant.

An urgent laparotomy was performed eight hours post presentation. The laparotomy revealed evidence of an ileal volvulus approximately 15cm in length with fibrotic adhesive bands at the apex. The twisted ileum appeared gangrenous and was resected with primary jejunio-ileal anastomosis done via a stapler. There was no evidence of congenital intestinal malrotation. The subsequent histopathologic examination of the resected specimen revealed the presence of an invasive carcinoid tumour located approximately midway through the length of the specimen.



Discussion

Aetiology

Small bowel volvulus is rare in an adult population. Incidence rates are relatively higher in Africa, Asia, the Middle East and India.^{3,4} The mortality associated with its sequelae is high and therefore increasing physician awareness of its diagnosis is crucial.

Small bowel volvulus can be attributed to primary or secondary causes. Small bowel volvulus without predisposing anatomical malrotation or anomalies is a rare incidence in adults. Primary volvulus is defined as volvulus occurring in the absence of anatomical defects whereas secondary volvulus is that occurring secondary to anatomical defects.² The theory behind primary small bowel volvulus occurring is based on strong abdominal muscles, high peristaltic tone as well as the presence of a bulky meal in the small bowel. The bulky bolus of food enters the proximal small bowel, causing the loop to descend inferiorly. This displaces empty small bowel loops upwards, initiating rotation of the mesentery and causing volvulus. Secondary volvulus has been often been attributed to malrotation, Meckel's diverticulum, leiomyomas of the mesentery, adhesive bands as well as malignancy.

Presenting complaint

The most common symptom associated with small bowel volvulus is acute onset severe abdominal pain with symptoms of bowel obstruction, nausea and vomiting.⁶

Diagnosis

The investigation of choice is a CT scan with sensitivity rates of 80%⁶ with the most characteristic finding of a whirlpool sign and the presence of gas indicating infarction. It has been reported in conditions such as adhesions, transverse colectomy as well as previous right hemicolectomy, however, it is not entirely specific.⁷ Another reported CT finding is triangular configuration of the bowel loops due to possible compression of the bowel loops secondary to the twist at the site of torsion.⁸ The 'beak' sign is also another reported sign on CT finding of tapering of three dilated bowel segments which appear analogous to a curved beak of a bird seen in barium enemas in studies of sigmoid volvulus.⁹

Frequently the plain abdominal radiograph will show multiple dilated small bowel loops with fluid levels consistent with small bowel obstruction. Rarely an abdominal radiograph can show dilated bowel loops with a 'spiral nebula' in the mid abdomen.¹⁰

However the physician should be aware of the false negatives and not neglect good clinical judgement based on clinical findings. This is evident in this particular case where investigations were non-specific and laboratory values not indicative of any pathological process.

Management

Some authors have recommended management of primary volvulus in the absence of gangrene as simple devolvulation. Some authors recommend resection and anastomosis in all small bowel volvulus in regardless of whether gangrene is present or not.^{4,11} In primary volvulus, fixation is recommended if resection is not performed with reports of recurrence in 30% of patients with simple devolvulation.⁶

Prognosis

Mortality has been reported to be extremely high ranging from 42.5 to 67%.¹²⁻¹⁵

Carcinoid Tumours

Carcinoid tumors of the small bowel represent approximately one-third of small intestine neoplasms and most commonly occur within 60 cm of ileocecal valve.¹⁶ Midgut carcinoid tumours are reported to be uncommon from an incidence of 0.5 to 1.5 per 100,000 individuals per year.^{17,18} Poor survival is associated with jejuno-ileal carcinoids with a five year survival rate of approximately 55%.¹⁹

Fibrosis is a well-documented sequelae, which occurs in carcinoid tumours. Many reports of retroperitoneal fibrosis, formation of bands as well as fixation of mesentery to the retroperitoneum have been reported.¹⁹ The extensive fibrosis can cause luminal obstruction, hydronephrosis and renal failure.²⁰

Currently, the exact pathophysiology behind the extensive fibrosis seen in some carcinoid tumours is unknown. Many theories have been postulated. The presence of 5HIAA especially with its associated with cardiac fibrosis have been documented.²¹⁻²³ The increased expression of PDGF-B on these tumours and synthesis of PDGF surrounding stromal components of these tumours contribute to the stimulation of connective tissue proliferation.²⁴ Other contributing growth factors include TGF-B known to be a potent stimulator of collagen synthesis.

In this particular case, the peculiar finding of fibrotic bands found at the apex of the volvulated loop, which could have led to the small bowel volvulus, can be explained due to the presence of carcinoid tumour.



Conclusion

To conclude, small bowel volvulus although rare should be a differential for any presentation of acute abdominal pain. There are no tests to exclude strangulated small bowel and laparotomy should not be delayed. Carcinoid tumours may also be associated with small bowel volvulus given its potential to cause fibrosis and the development of bands.

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PEER REVIEW

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CONFLICTS OF INTEREST

None

PATIENT CONSENT

The author Leong Chee Weng, declares that:

1. He has 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.