Appendiceal mucocoele with midgut malrotation

D Yap, J Hassall, GL Williams, ES McKain

Royal Gwent Hospital, Newport, UK

ABSTRACT

INTRODUCTION Malrotation of the midgut and appendiceal mucocoele are both extremely rare pathological conditions in adults. To our knowledge, there are only two reported cases in the English literature with a combination of both conditions.

CASE HISTORY A 65-year-old man presented with a 10-day history of upper abdominal pain associated with abdominal bloating and weight loss. He was otherwise fit and healthy with no significant past medical history. On examination, his abdomen was soft with tenderness and palpable fullness over the left upper quadrant. The initial blood test, chest x-ray and abdominal x-ray demonstrated no significant abnormality. Computed tomography showed a 17cm x 8cm x 6cm elongated cystic mass with possible malrotation of the intestines. Histopathology showed a low grade mucinous tumour of the appendix. At 12 months following surgery, there was no evidence of recurrence or postoperative complications and the patient was discharged from the care of the colorectal team.

CONCLUSIONS We report a patient with a combination of two rare conditions. This case illustrates how a combination of pathologies can present a challenge to the unwary general surgeon.

KEYWORDS

Abdominal pain – Intestinal malrotation – Mucocoele – Appendix

Accepted 2 May 2016

CORRESPONDENCE TO

Darren Yap, E: darrenyap.jx@gmail.com

Malrotation of the midgut and appendiceal mucocoele are rare pathological conditions in adults. A combination of both conditions would be clinically challenging as patients present with atypical signs and symptoms. The difficulty in diagnosing this uncommon combination can lead to possible delay in treatment. We report a case of an appendiceal mucocoele with a co-incidental finding of midgut malrotation. To our knowledge, there are only two previously reported cases in the English literature where both of these rare conditions present together (Table 1).^{1,2}

Case history

A 65-year-old man presented with a 10-day history of upper abdominal pain associated with abdominal bloating and weight loss. The pain was made worse with eating and drinking. There was no associated nausea, vomiting, change in bowel habit, fever, jaundice or lower urinary tract symptoms. He was an otherwise fit and healthy man with no significant past medical history. On examination, his abdomen was soft with tenderness and palpable fullness over the left upper quadrant. The mass did not extend down to the right iliac fossa.

On admission, the patient had a chest x-ray and an abdominal x-ray, which showed no significant abnormality. Laboratory results showed no abnormal findings other than raised C-reactive protein (118mg/l). His amylase and liver enzymes were normal.

The patient subsequently underwent computed tomography (CT), which showed a 17cm × 8cm × 6cm elongated cystic mass in the centre of the abdomen abutting the inferior surface of the transverse colon and superior margin of the urinary bladder. The lesion contained a large internal cyst, which had no specific diagnostic features (Fig 1). It was not tethered to any organ apart from the caecum. The radiologist commented that there may be a malrotation of the intestines (Fig 2).

Owing to ongoing symptoms and uncertain diagnosis, a laparotomy was performed during which the patient was found to have a malrotated caecum with a fluid filled mass at the tip of the caecum in place of the appendix. The caecum and mass were in the left upper quadrant (Figs 3 and 4). The mass was surrounded by a pseudocapsule with adhesions with sigmoid colon, descending colon, mesentery and small intestine. The lesion was resected and the ileocaecal junction preserved. There was no spillage of mucus or breach of the capsule. There was indeed malrotation of the small bowel and right hemicolon.

The patient recovered well from his surgery with no postoperative complications. Histopathology showed a low grade mucinous tumour (mucocoele) measuring 17.5cm × 7cm × 5cm, which was completely excised. An outpatient colonoscopy was performed, which was normal. CT at 12 months was also normal with no recurrence. There was no evidence of any recurrence or late postoperative complications and the patient was discharged from the care of the colorectal team.

Table 1 Summary of reported cases of combination of malrotation and appendiceal mucocoele						
Authors	Age / sex	Location	Size	Symptoms	Follow-up duration	Recurrence
Sato, 2001 ¹	76 F	Left lower quadrant	4cm x 4cm	Abdominal pain	23 months	No
Kawashima, 2001 ²	51 M	Lower abdomen	6cm	Abdominal pain	Not available	Not available
Present study, 2016	65 M	Left upper quadrant	17cm x 8cm x 6cm	Abdominal bloating and weight loss	2 years	No

Figure 1 Coronal and sagittal computed tomography showing appendiceal mucocoele

Figure 2 Axial computed tomography showing inversion of superior mesenteric artery (white arrow) and superior mesenteric vein (blue arrow)

Discussion

Mucocoele of the appendix is uncommon and found in 0.2–0.3% of appendix specimens, contributing only 8% of all appendiceal neoplasms.^{1,2,4} These lesions are typically identified in middle aged patients, with no definitive sex predilection.⁴ Appendiceal mucocoele is a descriptive term indicating

Figure 3 Appendiceal mucocoele in left upper quadrant

the appendiceal lumen is grossly distended with an abnormal intraluminal accumulation of mucus.^{1,5}

The aetiology is secondary to appendiceal obstruction from numerous possible causes such as a retention cyst, a simple mucocoele, mucosal hyperplasia, a mucinous cystadenoma and a mucinous cystadenocarcinoma.^{1,3,4} Other obstructing lesions such as adenocarcinomas of the colon, carcinoid tumours and endometriosis can also produce mucocoeles.⁴ Pathologically, it is believed that mucocoeles exist along a spectrum, beginning with benign appendiceal hyperplasia, leading to adenoma and finally to malignant transformation such as cystadenocarcinoma, similar to the progression of hyperplastic colonic polyps to colon adenocarcinoma.^{3,4}

The diagnosis can be made radiologically by computed tomography. An appendiceal mucocoele appears as a well circumscribed, low attenuation, spherical or tubular mass contiguous to the base of the caecum. Most mucocoeles are incidental findings; however, a small portion become symptomatic owing to increasing size, bleeding, infection, torsion or perforation.^{3,4}

During surgical excision, surgeons must take great care not to spill the abdominal mucocoele’s contents as morbidity

Figure 4 Mucocoele attachment to caecum in left upper quadrant

and mortality from pseudomyxoma are significant. Pseudomyxoma peritonei is a dreaded complication of mucocoele rupture, resulting in the seeding of mucin secreting epithelial cells into the peritoneal cavity, which causes gelatinous ascites.⁴

At present, there only two reported cases in the English literature of presentation with appendiceal mucocoele in adults and malrotation of the midgut. Intestinal malrotation is a congenital anomaly of the midgut. Between the 4th and 11th weeks of intrauterine life, the midgut undergoes a complex process of herniation out of the abdominal cavity and subsequent lateral fixation. Disturbance to this embryological process can result in various abnormalities, the most common of which is malrotation.²

This is detected in 0.2–0.5% of the adult population but it is only clinically significant in a minority as most adults remain asymptomatic throughout life.^{2,5} Nevertheless, some may suffer from chronic and unexplained abdominal

discomfort. They are often diagnosed as a result of an unexpected finding during abdominal surgery for other conditions or acute abdominal emergency situations. Rotational anomalies are simply an incidental finding; it is generally wise to treat the immediate problem and leave the anomaly alone.¹

The CT (Fig 2) demonstrates a number of the key features of intestinal malrotation. The third part of the duodenum fails to cross the midline so that the duodenojejunal junction is positioned abnormally on the right side below the duodenal bulb. There is inversion of the typical superior mesenteric artery (SMA)/superior mesenteric vein (SMV) relationship with the SMA on the right and the SMV on the left. The large bowel is predominantly situated on the left side of the abdomen and the small bowel on the right.

Unlike in children, intestinal malrotation in adults is usually of little clinical significance. However, owing to variation in anatomical positions, common pathologies may be present in unexpected locations, such as in our case.

Conclusions

We report a clinically challenging diagnosis as the patient presented with a combination of two rare conditions. CT is useful in helping the surgeons to plan a safe operative course. This case illustrates how a combination of pathologies can present a challenge to the unwary general surgeon.

References

1. Sato H, Fujisaki M, Takahashi T *et al.* Mucinous cystadenocarcinoma in the appendix in a patient with nonrotation: report of a case. *Surg Today* 2001; **31**: 1,012–1,015.
2. Kawashima K, Ishihara S, Amano K *et al.* Nonrotation of the midgut with appendiceal mucocoele in an adult. *J Gastroenterol* 2001; **36**: 44–47.
3. Mellado JM, Martín J, Solanas S *et al.* Uncommon causes of acute abdominal pain: multidetector computed tomography pearls and pitfalls for the radiologist on call. *Curr Probl Diagn Radiol* 2012; **41**: 179–188.
4. Arrington D, Jewett B, Sterner S *et al.* Incidental mucocoele of the appendix in a 15-year-old girl. *Pediatr Emerg Care* 2014; **30**: 555–557.