Unusual presentation of more common disease/injury

Appendiceal mucinous adenocarcinoma presenting as an enterocutaneous fistula in an incisional hernia

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Summary

A 68-year-old woman with a history of bone-graft harvesting from the right iliac crest presented with an incisional hernia and abscess at the graft donor site. Following incision and drainage of the abscess, CT demonstrated an enterocutaneous fistula between the appendix and bone-graft incision with appendicitis assumed to be the original cause of the abscess. At laparoscopy, the appendix was adherent to the hernia sac with mucinous material at the superficial orifice of the fistula site but not in the peritoneal cavity. Laparoscopic appendicectomy with fistula track excision was performed. Histological evaluation confirmed a well-to-moderately differentiated mucinous adenocarcinoma arising on a background of dysplastic villous adenoma. Tumour extended along the fistula track to involve the surface skin. A laparoscopic right hemicolectomy, lymph node dissection and wide local excision of the fistula track were carried out at a second procedure. Final histology confirmed pT4N1 tumour with clear resection margins.

BACKGROUND

Appendiceal mucinous adenocarcinoma is rare with most cases presenting as acute appendicitis. These tumours show a propensity to form fistulae to adjacent viscera, retroperitoneal tissues or to the skin surface. We report an unusual case of appendiceal mucinous adenocarcinoma presenting as an abdominal wall abscess over an incisional hernia. Following incision and drainage of the abscess cavity a discharging enterocutaneous fistula developed from the appendix to the incision site. This case demonstrates the ability of mucinous adenocarcinoma to form fistulae and underlines the importance of considering rare diagnoses when presented with abdominal wall abscesses.

CASE PRESENTATION

A 68-year-old woman presented to hospital in March 2009 with a hernia and abscess over a previous right iliac crest bone-graft site. She had previously been under the care of the orthopaedic surgeons at the same hospital having undergone a right-sided total knee replacement in March 2002 for osteoarthritis. Two years after her initial surgery the knee replacement became infected and was subsequently managed with a three-stage revision involving bone grafts from the right iliac crest in September 2005. The incisional hernia had been present over the right iliac crest site since that time and, although the patient had noticed the swelling, it had been asymptomatic. She had experienced no change in bowel habit and was otherwise well. A MRI scan of her pelvis ruled out osteomyelitis in the iliac wing, but showed bowel entering the hernia sac.

The abscess was managed with incision and drainage the following day. A large amount of seropurulent fluid was drained and omentum was noted to be visible through the musculature of the abdominal wall. The musculature was closed over the omentum and the wound left open with a

vacuum dressing applied. Three days later the patient was taken back to theatre; the wound was explored, washed out and partially closed. Initial culture results from the abscess showed growth of Lancefield group-G β -haemolytic streptococci sensitive to penicillin and erythromycin.

The patient had a persistent discharging sinus in the right loin, which she managed with a stoma appliance. She remained systemically well with a regular bowel habit and no faeculant discharge from the sinus.

INVESTIGATIONS

CT of the patient's abdomen and pelvis in June 2009 demonstrated a hernia of the transversus abdominis muscle with the appendix and omentum passing into the hernia. Importantly, the CT demonstrated a fistula between the appendix and the skin surface (figure 1). The fistula track between the appendix and skin surface was thought due to probable previous acute appendicitis or possibly an appendiceal tumour. Colonoscopy was carried out to exclude caecal or appendiceal base malignancy and was normal.

DIFFERENTIAL DIAGNOSIS

The differential diagnosis following CT of the patient's abdomen included a discharging sinus and abscess secondary to an episode of acute appendicitis. Tumours of the appendix, being rare, were lower on the differential list. As is commonly reported in most case series of appendiceal tumours, the diagnosis was not clear even at laparoscopy. Histological diagnosis remains crucial in identifying these tumours. Given the presence of mucin at the enterocutaneous fistula site at laparoscopy, the most likely neoplastic diagnosis was mucinous adenocarcinoma. Other tumours, such as carcinoid and colonic-type adenocarcinoma, could not be ruled out clinically.

Figure 1 Contrast-enhanced CT demonstrating the appendix (asterisk) passing into the hernia sac at the right iliac crest bone-graft donor site. The fistula track between the appendix and the skin surface is also clearly visible (arrows).

Figure 2 Photograph taken at laparoscopy showing the appendix entering and adherent to the abdominal wall defect. The adherent omentum has been retracted from the area superior and to the right of the appendix tip.

Figure 3 (A) Moderately dysplastic villous adenoma arising in the appendix. (B) Moderately differentiated mucinous adenocarcinoma extending along fistula tract to skin surface.

TREATMENT

In August 2009 the patient underwent a laparoscopic appendicectomy and excision of the fistula track. At operation, the appendix was found to enter and be adherent to the hernia sac with mucinous fluid at the external opening of the entero-cutaneous fistula site (figure 2). A laparoscopic appendicectomy and wide local excision of the enterocutaneous fistula was performed. The appendix and track were retrieved via the external opening of the track, and not via

the laparoscopic approach, specifically to avoid seeding mucin into the peritoneal cavity. Mucus was not sent for histology as it was felt likely that the diagnosis would be confirmed histologically.

Microscopic examination showed a well-to-moderately differentiated mucinous adenocarcinoma arising in the appendix on a background of a dysplastic villous adenoma (figure 3A). Tumour cells had invaded through the appendiceal wall and extended along the enterocutaneous fistula

track into the surface keratinising squamous epithelium (figure 3B). All resection margins, apart from the base of the appendix, were positive for tumour cells. On the basis of these histological findings, the patient was listed for a right hemicolectomy, lymph node dissection and wide local excision of the fistula track.

During September 2009 the patient returned to theatre for a laparoscopic right hemicolectomy. At operation, mucinous deposits were noted near to the appendiceal stump. Although there was no obvious widespread peritoneal mucin deposition, metastatic tumour spread was suspected to a single enlarged lymph node identified in the mesentery (video 1). This was excised en bloc with the right colon and arterial high-tie. The right hemi-colon was removed, along with a short (50 mm) segment of terminal ileum, and an anastomosis was fashioned between the ileum and proximal transverse colon. At the same operation, the patient also underwent wide local excision of the fistula track. A large amount of subcutaneous mucinous material was excised and the fistula track dissected to the peritoneum. The abdominal wall defect was closed with the aid of mesh.

Video 1 Laparoscopic lymph node dissection and arterial high-tie. 10.1136/bcr.11.2009.2472v1

Surgical specimens were submitted for histological examination. The hemicolectomy specimen showed microscopic evidence of signet ring mucinous adenocarcinoma with extensive lympho-vascular invasion. Although the resection margins were clear of tumour cells, two out of nine lymph nodes (including the apical node) contained metastatic tumour deposits. The extraperitoneal fistula track excision specimen demonstrated a cystic cavity containing malignant cells and mucin, lined by fibroadipose tissue, which was at least 7 mm clear of all resection margins.

OUTCOME AND FOLLOW-UP

The patient's postoperative recovery was unremarkable and she remains systemically well. She was referred to oncology for consideration of adjuvant chemotherapy. Postoperative CT demonstrated small-volume lymph nodes in the mesentery and right inguinal region. The patient has completed oxaliplatin and 5-FU chemotherapy and is under a surveillance programme for early detection of both recurrence and the possibility of peritoneal carcinomatosis.

DISCUSSION

Primary appendiceal tumours are rare. They represent 0.2–0.5% of all intestinal malignancies;^{1 2} only 4–6% of primary appendiceal neoplasms are adenocarcinomas and the vast majority are neuro-endocrine tumours.^{1–5} There are two main histological classifications of appendiceal adenocarcinoma: mucinous and colonic. The presenting signs and symptoms of appendiceal mucinous adenocarcinoma are usually those of acute appendicitis or a palpable abdominal mass⁶; however, unusual presentations include peritoneal carcinomatosis, haematuria due to bladder infiltration, direct invasion of the ascending colon noted on colonoscopy, hydronephrosis, retroperitoneal abscess, vaginal bleeding and cutaneous infiltration.^{7–15} These unusual presentations result from the propensity of appendiceal mucinous

adenocarcinoma to perforate and to form fistulae to adjacent structures.

Adenocarcinoma of the appendix is the most frequently perforating tumour of the gastrointestinal tract.¹⁶ This is thought to be due to anatomical peculiarities of the appendix, which has an extremely thin subserosal and peritoneal coat and the thinnest muscle layers of the whole gastrointestinal tract. Perforation of an appendiceal mucinous adenocarcinoma can result in widespread seeding of tumour cells and mucin deposits throughout the peritoneum with the development of peritoneal carcinomatosis in some, but not all, cases.^{1 15} This slowly progressive tumour with associated mucinous ascites significantly worsens the prognosis of mucinous adenocarcinoma,^{15 17–19} although this is disputed by one large case series.⁶ Recent evidence suggests that even small localised peri-appendiceal tumour deposits impact negatively on long-term survival in the absence of peritoneal carcinomatosis.²⁰ There was no evidence of perforation or of disseminated peritoneal mucin deposits or mucinous ascites in our patient either on CT or at laparoscopy. However, localised mucin deposition near to the appendiceal stump was seen at the second laparoscopy.

In addition to the risk of perforation, mucinous adenocarcinoma is thought to have a particular tendency for fistula formation.²¹ Many of the unusual presentations reported for primary appendiceal mucinous tumours are the result of fistula formation into adjacent viscera such as the urinary bladder, bowel and vagina as well as extra-peritoneally into the retro-peritoneal tissues or directly to the skin surface. The patient reported here developed an enterocutaneous fistula following incision and drainage of an extra-peritoneal abscess. We are aware of only five other case reports in the literature describing cutaneous fistulae arising from appendiceal mucinous cystadenocarcinoma.^{11 12 22–24} Fistulae in these cases arose spontaneously in locally advanced tumours, all of which had also spread into the ascending colon. In our case it is likely that the initial presentation with an abscess represented an infected mucocele, which may have discharged spontaneously through the skin surface if treated conservatively. Previous authors have suggested that such extra-peritoneal rupture of an appendiceal mucocele offers a favourable prognosis by preventing the development of peritoneal carcinomatosis.^{11 12}

Our patient initially underwent a laparoscopic appendicectomy. Following the histological diagnosis she was taken back to theatre for a right hemicolectomy and lymph node clearance. Nitecki *et al*⁶ reported one of the largest case series of appendiceal adenocarcinomas and advocate right hemicolectomy for all patients with mucinous adenocarcinoma. Their data show a survival advantage of hemicolectomy over appendicectomy alone (5-year survival, 73% vs 44%, $p < 0.01$). Hemicolectomy performed as a second procedure following appendicectomy (as in our patient) resulted in an up-staging of 38% of tumours. Nitecki *et al* also reported synchronous second primary malignancies in a large proportion (35%) of patients and suggest close surveillance for synchronous or metachronous tumours, particularly in the gastrointestinal tract. A later meta-analysis of 284 patients with appendiceal mucinous adenocarcinoma also showed a significant survival advantage of right hemicolectomy over appendicectomy.²⁵ Sugarbaker has

demonstrated increased survival in patients undergoing hemicolectomy as a single operation compared to those undergoing a staged procedure for appendiceal mucinous adenocarcinoma.²⁶ Current American College of Surgeons' guidelines recommend right hemicolectomy for all non-carcinoid appendiceal tumours with appendicectomy alone reserved for carcinoid tumours less than 1 cm in diameter.²⁷

Learning points

- ▶ Appendiceal tumours can present with unusual signs and symptoms related to the propensity of mucinous tumours to form fistulae.
- ▶ Right hemicolectomy is required to treat and stage appendiceal adenocarcinoma.
- ▶ Mucinous adenocarcinoma carries a risk of developing peritoneal carcinomatosis.
- ▶ The presence of mucin in association with appendicitis should lead the operating surgeon to suspect adenocarcinoma. In such cases it is important to avoid contamination of the peritoneal cavity with mucin. We also suggest obtaining a cytological analysis of the mucin as the presence of carcinoma cells confirms appendiceal adenocarcinoma rather than pseudomyxoma peritonei.
- ▶ Extra-peritoneal rupture of mucinous tumours may offer a more favourable prognosis by preventing the development of peritoneal carcinomatosis.

Competing interests None.

Patient consent Obtained.

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