Incidental mucinous cystadenocarcinoma of the appendix

J Lee, KR Gardiner, BG Wilson

Summary

The incidental finding of a rare appendiceal tumour in a previously healthy male patient is described. He presented as an emergency with a self-inflicted abdominal stab injury. At laparotomy, there was no visceral damage but the appendix was hugely distended and appendectomy was performed. Subsequent histological examination of the appendix revealed a low-grade mucinous cystadenocarcinoma. A right hemicolectomy was performed two months later which demonstrated no residual tumour.

Keywords: appendiceal tumour, cystadenocarcinoma

Appendiceal malignancies are rare and account for less than 0.5% of all intestinal tumours.¹ Diagnosis prior to surgery is difficult and rarely achieved. Controversies exist regarding their pathogenesis, management and prognosis as no single centre has accumulated a large experience. We report on a case of mucinous cystadenocarcinoma of the appendix with a most unusual presentation.

Case report

A 34-year-old man was brought by ambulance to the accident and emergency department at 01:41 hours with an abdominal stab wound. He described the mechanism of injury as due to a fall in his own home while holding a kitchen knife. On examination, he was intoxicated with alcohol and haemodynamically stable (pulse 95 beats/min and blood pressure 140/80 mmHg). A knife handle protruded from a wound in the left iliac fossa. The abdomen was soft to palpitation and normal bowel sounds were heard. All lower limb pulses were present and equal. Plain abdominal radiography showed a 15 cm long blade pointing towards the pelvis. There were no other abnormal findings. A urethral catheter was inserted and urinalysis was normal.

Laparotomy was performed via a lower mid-line incision. Although the knife breached the peritoneum, there was no injury to the viscera. However, a tensely distended appendix measuring 10 × 8 cm with a narrow base was observed. There was no evidence of mesenteric lymphadenopathy, peritoneal seedlings or hepatic metastases. Appendectomy was carried out with care being taken to avoid rupture of

the suspected mucocele. Postoperatively, the patient developed cellulitis of the stab wound which responded well to oral ampicillin. Recovery was otherwise uneventful and the patient was discharged on the 12th post-operative day.

Histological examination of the appendix showed mucin-secreting epithelium with focal papilliferous projections into the lumen and heaping of the nuclei. Mitosis was fairly prominent. A histopathological diagnosis of low-grade mucinous cystadenocarcinoma was made (figure). Although there was no evidence of local infiltration, there was mucosal ulceration and associated inflammation at the limit of excision.

The diagnosis was discussed with the patient and he decided to undergo further surgery to remove the appendix base and adjacent caecum. Two months later, the patient underwent an elective right hemicolectomy with end-to-end ileo-colic anastomosis with a single-layer of interrupted sutures. Post-operative recovery was uneventful and the patient was discharged after seven days. There was no evidence of residual tumour in the resected right hemicolon on histological examination.

Discussion

Mucocele of the appendix was first recognised as a pathological entity by Rokitansky in 1842.² Tumours of the appendix are rare and account for 0.5% of all intestinal tumours. They are associated with a second malignancy in the gastrointestinal tract in 15–20% of cases.³ Appendiceal tumours have a slight male preponderance with a ratio of 1.4:1. Mucinous

Figure Microscopic appearance of the appendiceal tumour (H&E)

Department of
Surgery, Mater
Infirmorum Hospital,
Belfast, Northern
Ireland, UK

J Lee
KR Gardiner
BG Wilson

Correspondence to
Jack Lee, Barbour Ward,
Royal Belfast Hospital for
Sick Children, Royal Trust
Hospitals, Grosvenor Road,
Belfast BT12 6BA, Northern
Ireland, UK

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Mucinous cystadenocarcinoma of the appendix: clinical features
• presents with appendicitis, palpable mass in the right iliac fossa, rectal bleeding, pain, fistula or pseudomyxoma peritonei   • may be diagnosed by plain abdominal X-rays, contrast enema examinations, ultra sound scan or computed tomography   • rarely spreads by the lymphatics   • slower growth rate than adenocarcinoma   • 50% of patients present with intra-abdominal metastasis or pseudomyxoma peritonei

Mucinous cystadenocarcinoma of the appendix: prognosis
• 65% of patients treated with right hemicolectomy survived 10 years   • patients with pseudomyxoma peritonei have a 50% 5-year survival rate   • good prognosis with early complete excision of the tumour

cystadenocarcinoma is the second most common appendiceal tumour and accounts for 8% of all primary appendiceal malignancies. The patients with mucinous cystadenocarcinoma are usually younger than the typical patient with adenocarcinoma of the appendix.³

Classical roentgenographic appearance of appendiceal mucinous tumour are: a sharply circumscribed globular soft tissue mass; medial displacement of the caecum; and calcium deposition in a soft tissue mass. On barium enema examination, the appendix fails to fill with contrast. Ultrasound scan may demonstrate the cystic nature of the mass and any exuberant mucosal proliferation. Computed tomography (CT) can be diagnostic for a mucinous appendiceal tumour by finding a mass compressing or adjacent to a contrast-filled caecum.⁴

As malignant mucocele of appendix rarely spreads via lymphatics, it has been suggested by Anderson and colleagues that appendicectomy is adequate⁵ but others recommend right hemicolectomy.^{3,4} Malignant mucocele of the appendix has a slower growth rate and a higher survival rate than is found with appendiceal adenocarcinoma. Stephenson and colleagues⁴ reviewed 53 appendiceal mucinous cystadenocarcinomas treated by either appendicectomy or right hemicolectomy. At 10 years, survival was 65% in patients treated with right hemicolectomy, in contrast to 37% among

Tumours of the appendix: summary points
• associated with a second malignancy in the gastro-intestinal tract in 15–20%   • slight male preponderance   • mucinous cystadenocarcinoma is second most common   • incidental finding of this tumour is uncommon   • excision of the tumour and right hemicolectomy is the appropriate management   • surgical removal of the tumour offers a cure if disease detected early   • radiotherapy and chemotherapy have not been found to be effective in disseminated disease

those who had undergone appendectomy. Rutledge *et al*³ found a 70% 5-year and 65% 10-year survival rate after right hemicolectomy. Even patients with pseudomyxoma peritonei have a 50% 5-year survival rate.³ Aggressive debulking, omentectomy and oophorectomy are all considered worthwhile for advanced disease.^{1,3,4} The development of pseudomyxoma peritonei often requires repeated surgery for intestinal obstruction or fistula formation. Radiotherapy and chemotherapy have not been found to be effective.^{4,6}

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