CASE REPORT

Two rare cases of appendiceal collision tumours involving an appendiceal mucinous neoplasm and carcinoid

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SUMMARY

We report two rare cases of appendiceal collision tumours, each involving an appendiceal mucinous neoplasm with a concomitant appendiceal carcinoid. The first case presented with acute appendicitis while the second presented with an incidental radiological finding of an enlarged appendix as part of evaluation for raised serum carcinoembryonic antigen. Diagnosis in both cases was made postoperatively on histological assessment following appendicectomy. Clinical and histological features of both tumour types in each case did not necessitate further surgical resection or adjuvant treatment. Both patients did well postoperatively and had no evidence of disease recurrence on follow-up.

BACKGROUND

Appendiceal collision tumours are extremely rare entities, with less than five reported cases within the existing literature. Examining the clinical features and management of such patients helps contribute to further understanding of the optimal clinical evaluation and management of such tumours.

CASE PRESENTATION

Case 1

A 59-year-old Chinese woman with no significant medical history of note was admitted for a 1-day history of right iliac fossa (RIF) pain associated with anorexia and vomiting. On physical examination, she was febrile with a temperature of 39.5°C and had RIF tenderness with guarding. Initial investigations were only notable for a raised total white blood cell count of 13.02 (reference range 4.0–10.0), suggestive of infection.

With a working diagnosis of acute appendicitis, the patient underwent a diagnostic laparoscopy, which revealed a suppurative inflamed appendix without perforation, associated with pus in the RIF and pelvis. Her surgery was converted to an open procedure, and she underwent an open appendicectomy uneventfully. She recovered well postoperatively and was discharged on postoperative day 3.

The final histology was an acutely inflamed appendix with a circumferential mucinous adenoma at the distal appendix as well as a small carcinoid measuring 2 mm in diameter at the tip (figure 1). At the patient's last follow-up, she was 5 years out of surgery, and a CT scan performed showed no signs of disease recurrence.

Case 2

A 52-year-old Chinese man with no significant medical or smoking history of note was referred to our centre in view of a persistently elevated serum carcinoembryonic antigen (CEA) level first detected on routine health screening 2 years prior. Initial endoscopic evaluation of the upper and lower gastrointestinal tract, and CT scan of the abdomen, performed at that time, did not reveal any significant abnormalities. He was subsequently placed on observation with serial trending of his CEA level. This revealed a progressively increasing trend (17.0–31.8 ng/mL over a 2-year period), which prompted the current referral to our tertiary centre.

All initial laboratory tests were within normal limits as well except for the elevated CEA. CT of the patient's abdomen and pelvis revealed a tubular cystic lesion measuring 3.4 cm attached to the caecum, likely appendiceal in origin, with no associated radiological evidence of metastatic disease.

Figure 1 Case 1 histology demonstrating mucinous adenoma on H&E stain (top panel) and carcinoid on immunohistochemical staining for CD56 (bottom panel) of the appendix.

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Colonoscopy was again repeated, and this did not reveal any mucosal abnormalities in the caecum to suggest intraluminal involvement.

The patient subsequently underwent a diagnostic laparoscopy, which revealed a 3.5 by 5.0 cm appendiceal tumour with involvement of neither the appendiceal base nor of the surrounding structures, and no obvious peritoneal disease. We then proceeded with a laparoscopic appendicectomy in the same setting. Intraoperative frozen section revealed a low-grade mucinous appendiceal neoplasm and, hence, a simple appendicectomy alone was deemed sufficient. The patient recovered uneventfully postoperatively and was discharged on postoperative day 2.

The final histology was a low-grade mucinous appendiceal tumour with an incidental finding of a distinct focus of carcinoid measuring 3 mm in maximum dimension (figures 2 and 3). All resection margins were clear, with no mesoappendiceal invasion. At present, this patient has been less than 3 months out of surgery, with a plan for a surveillance CT scan of his abdomen and pelvis in 6 months.

DISCUSSION

Primary appendiceal neoplasms are uncommon clinical entities usually discovered incidentally following an emergent appendicectomy for appendicitis in approximately 5% of cases.¹ True appendiceal collision tumours, defined as the presence of two histologically distinct tumour types of different clonal origins within the appendix with no transition between them, are even rarer, with less than five cases reported in the literature thus far.^{2–4}

Figure 2 Case 2 gross specimen demonstrating carcinoid in section near tip of appendix (top panel) and multiple sections containing a mucinous neoplasm of the appendix (bottom panel).

Figure 3 Case 2 histology demonstrating mucinous adenoma on H&E stain (top panel) and carcinoid on immunohistochemical staining for CD56 (bottom panel) of the appendix.

In these two rare cases of appendiceal collision tumours we report, it is evident that a spectrum of clinical presentations exists, from an asymptomatic patient with an incidentally noted elevated CEA to an acute presentation of appendicitis. Of note, both patients were aged 50 years and above, highlighting the importance of considering the presence of an appendiceal neoplasm with increasing age.⁵

Following the postoperative diagnosis of appendiceal collision tumours in both cases, no further management was deemed necessary after taking into consideration optimal management of each histological tumour type found. Appendiceal mucinous neoplasms constitute a histological spectrum including mucinous adenomas, low-grade mucinous neoplasms and mucinous adenocarcinomas.⁶ Management of the first two entities (seen in each of our two cases, respectively) simply requires an appendicectomy with clear margins.⁷ Appendiceal carcinoids can similarly be managed with a simple appendicectomy with clear margins provided the tumour size is less than 2 cm and there is no evidence of mesoappendiceal invasion.⁸ Both patients did well postoperatively with no evidence of disease recurrence thus far on follow-up.

Further, as evidenced by our second case, a laparoscopic approach appears to be a safe and feasible option for selected appendiceal tumours, with similar findings reported in the literature for both, appendiceal carcinoids⁹ and appendiceal mucinous neoplasms.¹⁰

Owing to the rarity of appendiceal collision tumours, there are no existing recommendations for an optimal follow-up regime postoperatively. We believe it is prudent to follow these patients up with regular interval imaging and tumour markers (eg, CEA for mucinous appendiceal neoplasms and chromogranin A for appendiceal carcinoids) if elevated preoperatively. Frequency of tests and duration of follow-up should be based on the histological tumour type with worse prognosis. In our two cases, neither the appendiceal mucinous neoplasm nor the appendiceal carcinoid have established guidelines for post-operative follow-up in the literature, with some reports suggesting that there is no routine need for any follow-up in view of the exceedingly low recurrence rates.⁸ As such, we believe that the follow-up regime ought to be tailored in discussion with the patient, weighing the risk of recurrence against the costs and cumulative radiation exposure of regular interval scans.

Learning points

- ▶ Appendiceal neoplasms are uncommon but clinically important considerations in patients with appendicitis or imaging appendiceal abnormalities.
- ▶ The management and postoperative follow-up of appendiceal collision tumours both require a combined consideration of the involved histological tumour types.
- ▶ Laparoscopic appendicectomy is a safe option for selected cases of appendiceal neoplasms.

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Contributors HLT drafted the manuscript, worked on subsequent revisions and liaised with the pathology department for the histological slides and gross specimen photos. GHCT worked on manuscript revisions. MT worked on manuscript revisions.

Competing interests None declared.

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