CASE REPORT

Appendiceal mucocele: a missed diagnosis

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SUMMARY

A 38-year-old man presented to clinic with recurrent right iliac fossa pain from a young age; three episodes of which required hospital admission. He was otherwise well with no associated symptoms. In view of persistent pain and recurrent admissions a CT scan was arranged which showed a dilated retrocaecal appendix suggesting recurrent appendicitis. Further to CT findings the patient underwent an urgent appendicectomy for a presumed diagnosis of chronic appendicitis but histology in fact revealed a cystadenoma of appendix. Postoperatively the patient's symptoms resolved and he was discharged from surgical care. It is important to note that in this case the diagnosis of appendiceal mucocele was not considered therefore not particularly accounted for during surgery. Has this been a malignant cystadenocarcinoma any spillage of malignant cells into the abdominal cavity during the operation could have a grave impact on long-term recovery and prognosis.

BACKGROUND

This case discusses a rare cause of a common symptom presenting in surgical patients. Chronic right iliac fossa pain is often dismissed and rare pathology can be missed. This symptom can occur in patients with gynaecological problems, ileocecal pathology, diverticular and appendicular pathology like appendiceal mucocele. The latter diagnosis is one which the clinician should be aware of, particularly in patients with longstanding history. This case illustrates the importance in assessing for an appendiceal mucocele and emphasises the positive implications that careful planning and early recognition can have on its management.

CASE PRESENTATION

A 38-year-old man was referred to a general surgical outpatient clinic with a history of recurrent episodes of right iliac fossa pain, colicky in nature. In total he had presented to hospital three times with the same problem, the first episode in 1999. The most recent episodes were in 2007 and 2009. On both occasions he was investigated and sent home within 2 days following conservative management. He was also seen multiple times by the general practitioner in between hospital admissions and sent home with reassurance and analgesia and the pain tended to self-resolve within 1–2 days. On further questioning, the patient admitted to a flare up of pain every 2–3 months but he only sought medical attention when it reached an intolerable level of severity.

The pain always localised around the umbilicus and right iliac fossa, sometimes radiating into the groin. On two occasions there was associated

nausea but no vomiting. His bowels functioned regularly and there were no urinary symptoms. There was no history of weight loss, appetite loss or fatigue. Family history of bowel pathology was unremarkable.

Following a further episode in 2011, he was referred by his general practitioner to urology with a possible diagnosis of renal colic. Examination of the abdomen, including the flanks and external genitalia was unremarkable with no palpable mass.

INVESTIGATIONS

On each admission bloods were normal except in 2007 where there was a slight increase in white cell count ($15.6 \times 10^9/l$). Urine dipsticks were always negative for leucocytes and nitrites and there was no evidence of bacterial growth when sent for culture and sensitivity.

Ultrasound scan in 1999 and 2007 did not reveal abnormalities. In 2011, no stones were visualised on his abdominal radiograph. In view of the prolonged nature of the symptoms, a CT scan of abdomen was performed to assess the problem further. The scan revealed a retrocaecal appendix which was slightly prominent measuring up to 16 mm in diameter (figure 1). Hence he was referred to the general surgeons on an urgent basis. In view of clinical presentation and CT findings a diagnosis of recurrent appendicitis was made and he was scheduled to have an urgent elective appendicectomy.

DIFFERENTIAL DIAGNOSIS

The history presents itself as a young healthy man with recurrence of an isolated symptom of right-sided abdominal pain. On one occasion there was a rise in inflammatory markers which indicates the possibility of intermittent inflammation; however, there were no other clinical markers to support this. The colicky nature of the problem indicated the possibility of calculi but he had few risk factors for gallstones and this was disproven twice on ultrasound scan. Abdominal radiograph was negative for renal calculi. There were no features of weight loss or change in bowel habit and no family history to support a possible diagnosis of inflammatory bowel disease or carcinoma. At each admission, appendicitis was queried; however, after a period of observation the patient symptoms resolved and the problem was put down to non-specific abdominal pain. Even as a definitive diagnosis was reached on CT scan and in surgery, it appeared that appendicular mucocele was never considered. This illustrates that it is typically a diagnosis that is missed and not considered as part of the differential for recurrent right-sided abdominal pain.

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Figure 1 CT scan abdomen showing dilated appendix.

TREATMENT

An elective laparoscopic appendicectomy was performed. At surgery the appendix was found to be dilated particularly at the base. Endoloops were used to remove the appendix with care taken to prevent spillage of appendiceal contents intraperitoneally with the help of a Bert bag. Histology samples showed mucinous cystadenoma of the appendix. There was no evidence of epithelial elements invading into underlying connective tissue stroma so malignancy was ruled out.

OUTCOME AND FOLLOW-UP

The patient was discharged the next day and seen in outpatient clinic 2 weeks later. In view of histology and negative findings on laparoscopic inspection of the abdomen, the patient was reassured and no plans were made for further investigation.

DISCUSSION

Mucocele of the appendix is defined as distention of the appendicular lumen due to accumulation of mucoïd substances.^{1 2} It is a rare lesion with very few cases reported in the literature and incidence quoted as low as 0.2–0.3%.^{1 2} The disease has been classified into four subgroups according to the characteristics of the epithelium³:

- ▶ Simple mucocoeles which result secondary to obstruction of appendicular flow;
- ▶ Mucocoeles with hyperplastic epithelium;
- ▶ Benign cystoadenomas which exhibit villous adenomatous changes and
- ▶ Malignant mucinous cystadenocarcinomas which exhibit glandular stromal invasion or the presence of epithelial cells in peritoneal implants.

Thicknesses of the wall of different subtypes of mucocoele vary but the lumen tends to be less distended in a simple, hyperplastic mucocoele when compared with a cystoadenoma or cystoadenocarcinoma in which luminal dilation can be up to 6 cm.³ The most common type of mucocoele is a cystoadenoma which occurs in 63–84% cases.^{3 4}

Diagnosis can be suggested by imaging and confirmed by surgery and histopathology of the specimen. There appears to be a higher incidence in females (4 : 1) with more cases presenting in patients above the age of 50.^{2 3} The cause for this remains unknown. Clinical presentation can vary from the patient being asymptomatic in 50% cases to the patient exhibiting symptoms that mimic those in acute appendicitis or appendicular mass. Tenderness in the right iliac fossa with a palpable mass are two most common presenting symptoms. The patient is usually generally well with no signs of haemodynamic compromise. In few patients there have been reports of weight loss,^{4 5} a change in bowel habit and bleeding per rectum.⁵ Whether these symptoms are secondary to the condition remains a subject of debate. There are usually no symptoms of guarding or rebound except where pseudomyxoma peritonei has occurred. This condition is secondary to spontaneous or iatrogenic rupture of the appendix.³

Diagnosis of an appendicular mucocoele can often be difficult due to the asymptomatic nature of the disease. In 50% cases the lesion is an incidental finding at the time of surgery.^{4 5} However if the diagnosis is borne in mind, diagnostic tests can be of great value particularly in planning for surgery. Careful resection can prevent perforation and spreading of the potentially malignant cells (malignant cystoadenocarcinomas) into the peritoneum which may have a dramatic impact on prognosis. In malignant mucocoeles the 5 year prognosis can fall from 90% to 25% as a result of complications of pseudomyxoma peritonei.³ Therefore, it is essential that

clinicians are aware of the disease when seeing a patient who presents with recurrent pain or a mass in the right iliac fossa.

Once the suspicion of a mucocoele is raised, a number of investigations can help to assess the likelihood of the diagnosis. This is mainly in the form of imaging. Ultrasonography demonstrates a cystic encapsulated lesion with the internal variable echogenicity related to the density of the mucus. However such findings can be attributed to other conditions such as fluid in a diverticulum, a diverticular or appendicular abscess, and in women gynaecological conditions such as ectopic pregnancy and salpingitis have been suspected.⁴ CT is more specific therefore can be imperative in the correct preoperative diagnosis. Typically, it demonstrates a hypoattenuated well-encapsulated smooth or lobulated mass. More complex masses tend to be associated with cystadenocarcinomas and may demonstrate infiltration into adjacent structures.⁴ In benign mucocoeles CT will more likely to show displacement of bowel loops secondary to mass effect.⁵ The absence of inflammatory changes helps to distinguish a mucocoele from acute appendicitis. However CT is not 100% specific as demonstrated by this case. There remains scepticism around whether or not recurrent appendicitis truly exists and some clinicians may have been doubtful over the CT report in this case. However there are data in the literature that suggests that up to 15% patients suffer with recurrent right iliac fossa pain which resolves spontaneously, prior to an episode which eventually requires an appendicectomy.⁶ Therefore, it is reasonable to suspect such a diagnosis.

Definitive management of an appendiceal mucocoele is surgical resection. However, there should be continues debate regarding the most appropriate approach and extent of resection required. Literature contradicts itself in one sense as it cautions against laparoscopy due to the risk of rupture.³ On the contrary, it accepts that laparoscopy provides an opportunity to evaluate the entire abdominal cavity for evidence of dissemination.⁵ Laparoscopy also has the advantage of a more rapid recovery and better cosmetic appearance than a laparotomy. Margins of resection should be considered according to the type of mucocoele. This is where sufficient preoperative investigations can help to advise the surgeon. In simple mucocoeles appendicectomy will suffice.³ In cystadenocarcinomas it is advisable to perform a right hemicolectomy as well as conduct a thorough inspection of the rest of the abdominal cavity due to the association between the appendicular mucocoele and other tumours, particularly of colonic or ovarian origin.⁴ However, recent data has suggested that there is actually no survival advantage in patients with cystadenocarcinoma undergoing right hemicolectomy.⁷ It is advised that this only becomes

necessary when there is local invasion in the bowel.⁷ Surgical management of a benign cystadenoma remains under debate. Some scholars advise simple appendicectomy if the mucocoele does not involve the caecum but others suggest that nevertheless, resection should be extended to the caecum in a broad-based mucocoele.¹ According to the limited cases reported there is no change in outcome when a benign cystadenoma is treated with minimal or extended resection.

Ultimately, mucocoele of the appendix is a condition of which all clinicians should be made aware. There is a broad spectrum of disease and a minority of cases can be potentially lethal. Therefore earlier recognition of the disease and appropriate planning can help to ensure a safer approach to resection.

Learning points

- ▶ Consider the condition of appendicular mucocoele when assessing a patient who presents with recurrent right iliac fossa pain.
- ▶ Careful surgical planning is required to avoid risk of pseudomyxoma peritonei.
- ▶ In cases where a cystadenocarcinoma is suspected always carefully inspect the abdomen for associated malignancies.

Competing interests None.

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