CASE REPORT

The appropriate management of an appendiceal mucocele

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SUMMARY

Appendicectomy is one of the most commonly performed surgical procedures. Appendiceal mucocele is a relative rare disease, but appropriate management is critical. Indeed, the intact removal of a mucocele represents a curative treatment; conversely, a rupture may result in the spread of epithelial cells throughout the peritoneal cavity (pseudomyxoma peritonei). We report a case of a 61-year-old woman, admitted to our department, who underwent resection of an appendiceal mucocele, focusing, in the discussion, on the clinical and surgical management of this disease.

BACKGROUND

Appendicectomy is one of the most commonly performed surgical procedures. Appendiceal mucocele has been reported to have an incidence of 0.2–0.3% in all appendix specimens, and is characterised by the accumulation of mucoid matter within the appendix lumen.^{1 2} Owing to the large number of appendicectomies performed each year, radiologists and surgeons should be familiar with this condition, since the appropriate management of this particular disease is critical.

The intact removal of a mucocele represents a curative treatment; conversely, a rupture, either spontaneous or accidental, may result in the spread of epithelial cells throughout the peritoneal cavity in the form of multiple mucinous deposits (pseudomyxoma peritonei).^{1 3} On the basis of this background, the aim of this manuscript is to report the case of an appendiceal mucocele and to review the clinical and surgical management of this disease.

CASE PRESENTATION

A 61-year-old woman presented to our department with a 4-month clinical history of mild abdominal pain (localised in the epigastrium), with referred right shoulder pain. She denied fever, urinary dysfunction or change in the intestinal transit. On admission, laboratory tests were within normal values and physical examination revealed no abnormalities.

INVESTIGATIONS

Plain chest X-ray film was negative for pulmonary and/or chest diseases. The patient underwent an abdominal ultrasound (US), which disclosed a cystic lesion in the right abdominal lower quadrant. Subsequently, the patient underwent a contrast-enhancement CT scan (figure 1), which documented a retrocecal mucoid mass, consistent

with a mucocele, with no signs of intraperitoneal ascites.

TREATMENT

The patient underwent appendicectomy via McBurney laparotomy. A giant retrocecal appendix was found and after adhesiotomy, a standard appendectomy was performed, removing the mass without spilling or rupture.

Intraoperative fresh frozen sections of the specimen were processed and resulted negative for neoplastic lesions.

OUTCOME AND FOLLOW-UP

Pathological examination was consistent with a mucocele associated with an appendiceal adenoma. The postoperative course was uneventful and the patient was discharged on postoperative day 4.

DISCUSSION

Clinical presentation of a mucocele could be non-specific and up to 50% of patients could be asymptomatic.¹ Therefore, radiological imaging is critical. US and CT scans are most often employed, usually documenting a well-encapsulated cystic mass in the right lower abdominal quadrant, often associated with mural calcifications.^{1 4} Differential diagnosis could be highly important, since a number of patients undergoing surgery may have a misdiagnosis of appendicitis; on this basis, Lien *et al* retrospectively analysed a case-matched series of patients with histologically confirmed mucoceles comparing with appendicitis. According to their results, an appendiceal diameter greater than 15 mm documented by sonography correlated with a mucocele with a sensitivity of 83% and a specificity of 92%.⁵ In line with these findings, and also comparing with appendicitis, Saylam *et al* recently confirmed that an outer diameter greater than 10 mm (by sonography) and a mean white body cell count (WBC) count lower than 11 000/mm³ were statistically correlated with a pathological diagnosis of mucocele. Interestingly, a significant number of patients with appendiceal mucocele had a positive microscopic haematuria comparing with appendicitis ($p=0.019$). On this basis, the authors assigned a point to each of these factors (large appendiceal diameter, low WBC count and presence of microscopic haematuria) resulting in a predictive score and a stratification of patients into three groups (Score I: 0 points; Score II: 1 points; Score III: ≥ 2 points). The scoring system for patients with ≥ 2 points was proven to have an area under the curve (AUC) of 0.855 using a receiver-operating

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Figure 1 Contrast-enhanced CT scans documenting a retrocecal appendiceal mucocoele, measuring 6×4 cm, with no signs of intraperitoneal ascites.

characteristic curve with a sensitivity of 73.33% and a specificity of 89.87%.⁶ Of note, the patient we presented had an appendiceal diameter of 40×60 mm (by CT scan), a WBC count of 9900/mm³ and a preoperative urine sample documenting 4 erythrocytes/μL.

Preoperative work up also discloses the presence of mucinous ascites in the pelvis, right abdominal quadrants, and between the liver and diaphragm, as a sign of rupture. According to Sugarbaker,³ there is a distinctive pattern of dissemination

within the peritoneal cavity, which follows the principles of fluid hydrodynamics and gravity distribution. Dhage-Ivatury recently proposed an algorithm for surgical management of a mucocoele. Patients should be referred for surgical removal, and in case of a non-perforated mucocoele (presenting with negative cytology, negative resection margins, with absence of appendiceal nodes) a standard appendicectomy should be performed, requiring no long-term follow-up.¹ Since it is very important to prevent rupture or spilling of mucoid material, an open laparotomy

or a conversion of a laparoscopic to a laparotomic procedure could be required. Also, according to Dhage-Ivatury and Sugarbaker, when a dysplastic mucocele has ruptured and mucinous carcinomatosis or pseudomixoma peritonei has been diagnosed, minimal surgery (including appendicectomy with free margins, appendiceal lymphadenectomy and sampling of the ascites) should be performed with the goal of establishing a diagnosis; in addition, the authors suggest irrigating the surgical incisions to minimise neoplastic seeding. Following and on the basis of the pathological and cytological reports, patients should be referred to a specialised centre for the further treatment and follow-up.¹ Treatment may range from appendicectomy to right colectomy and cytoreductive surgery, heated intraoperative intra-

peritoneal chemotherapy and early postoperative intraperitoneal chemotherapy, on the basis of presence of neoplastic cells in the (1) mucinous ascites, (2) resections margins and/or (3) appendiceal nodes.¹ Even though some controversies still exist regarding the clinical and histopathological definition of the pseudomixoma peritonei syndrome, this condition requires a multidisciplinary approach in a referral centre, since these treatments are affected by a high morbidity rate; 5-year survival rates have been reported ranging from 50–96%.¹ Ronnet *et al*⁷ classified mucinous appendiceal neoplasms on the basis of prognosis as: disseminated peritoneal adenomucinosis (least aggressive) and peritoneal mucinous adenocarcinoma (most aggressive). According to Sugarbaker, patients with a less aggressive disease treated by cytoreductive surgery plus perioperative intraperitoneal chemotherapy and achieving a complete cytoreduction, have a 70% survival rate at 20 years.⁸

Learning points

- ▶ Preoperative imaging should focus on the mucocele and on the presence of ascites as a sign of rupture.
- ▶ In case of a non-perforated mucocele (presenting with negative cytology, negative resection margins, with absence of appendiceal nodes), a standard appendicectomy should be performed.
- ▶ According to Sugarbaker, when a dysplastic mucocele has ruptured and mucinous carcinomatosis or pseudomixoma peritonei has been diagnosed, surgery should be performed with the goal of establishing a diagnosis; following this (on the basis of the pathological and cytological reports), patients should be referred to a specialised centre for further treatment and follow-up.
- ▶ Treatment may range from appendicectomy to right colectomy and cytoreductive surgery, heated intraoperative intraperitoneal chemotherapy and early postoperative intraperitoneal chemotherapy according to specific features including the presence of neoplastic cells in the mucinous ascites, resection margins or nodes.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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