Case report

Case of intraperitoneal sepsis secondary to rupture of the appendix on the background of pseudomyxoma peritonei

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HIGHLIGHTS

- Intraperitoneal sepsis secondary to appendiceal rupture is rare.
- Surgeons may face an emergency of intraperitoneal sepsis before the planned surgery or as a primary presentation.
- With combined therapy, the prognosis of mucinous appendiceal adenoma is excellent.

ABSTRACT

Introduction: Pseudomyxoma peritonei (PMP) is characterised by gelatinous ascites and pools of mucin associated with neoplastic mucinous epithelium within the peritoneal cavity. It can rarely present as acute intraperitoneal sepsis, requiring urgent medical attention.

Presentation of case: A 59-year-old male was referred to our centre in February 2014 following a diagnostic laparotomy, which showed jelly-like material with occasional epithelial cells. He was listed for peritonectomy in a month's time at our centre. Three weeks later, he was admitted urgently to our hospital due to generalised abdominal pain and watery diarrhoea. Examination at admission was unremarkable. On the following day, he became haemodynamically unstable and was suspected to have intraperitoneal sepsis due to infected PMP. At emergency laparotomy, we found gross intraperitoneal sepsis and did extensive debulking of tumour, appendectomy and extensive division of adhesions. Another laparotomy was done 24 h later for washout. He was discharged three weeks after.

Discussion: Although we have done 780 peritonectomy procedures, this was the first patient with this presentation of widespread intraperitoneal sepsis. Continuous mucous production of appendiceal adenoma can lead to appendiceal rupture. The appendix may decompress by perforation and then reseal. However, one episode of appendiceal rupture can cause bacterial contamination of PMP, leading to sepsis.

Conclusion: Intraperitoneal sepsis secondary to appendiceal rupture is rare. Hence surgeons may face an emergency of intraperitoneal sepsis during waiting period of planned CRS or as a primary presentation. With combined therapy of CRS and PIC, the prognosis of mucinous appendiceal adenoma is excellent.

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1. Introduction

Pseudomyxoma peritonei (PMP) is a slowly progressive condition characterised by gelatinous ascites and pools of mucin associated with neoplastic mucinous epithelium within the peritoneal cavity [1]. It is also a pathologic diagnostic term for a group of mucinous tumours derived from different sites that share the common characteristics of abundant extracellular mucin. Based on clinicopathological features of 109 cases, Ronnett et al. classified PMP into three groups, namely, disseminated peritoneal adenomucinosis (DPAM) and peritoneal mucinous carcinomatosis (PMCA) and hybrid type [2]. Peritoneal lesions of DPAM demonstrate scant simple to low-grade adenomatous epithelium, in contrast to histological features of mucinous carcinoma in the lesions of PMCA. Hybrid type refers to patients with mixed features of DPAM and PMCA. It is further divided into PMCA-I and PMCA-D. PMCA-I refers to peritoneal lesions which mainly show features of DPAM but also contained areas of well-differentiated mucinous adenocarcinomas. In contrast, PMCA-D contains poorly differentiated lesions similar to PMCA. This pathological classification is important for guiding therapeutic approaches and predicting...
prognosis. PMCA is associated with more aggressive behaviours and histological evidence of mitotic activity and cytologic atypia in contrast to little histological evidence of malignancy in DPAM. Thus it is currently recommended to add chemotherapeutic agents to therapeutic regimen for patients with PMCA. Also, DPAM is associated with a significantly more favourable prognosis than PMCA [2]. PMCA-I/D have similar prognosis with PMCA.

Although DPAM has a slow growing nature and is considered as benign, its acute presentation with sepsis can occur but appears to be very rare [3]. It is important for surgeons to consider possibility of intraperitoneal sepsis for patients with a known background of PMP especially for those who are haemodynamically unstable.

2. Presentation of case

A 59-year-old male presented with one-week history of acute generalised abdominal pain and four-day history of watery diarrhoea in March 2014 on background of PMP diagnosed in February 2014, who was waiting for workup and surgery at our centre. The pain was alternating between dull and sharp in nature and taking a few hours to go away slowly. He also complained of watery diarrhoea. The stool was dark brown without blood. It was estimated to be 12 times per day and persisted when fasting. Associated symptoms included loss of appetite, sweating, generalised aches, mild breathlessness and dry cough.

His PMP was diagnosed in February 2014, following an episode of bowel obstruction. He presented with sudden onset of abdominal pain and a fever of 38 °C to a regional hospital in February. Computed tomography (CT) showed free fluid and peritoneal nodules. His blood results showed C-reactive protein (CRP) of 300 mg/L, white cell count (WCC) of 17 × 10^9/L, CEA of 7.2 ng/ml and CA19-9 of 68 U/ml. Subsequently he underwent a diagnostic laparotomy at the regional hospital. The material removed was reported as jelly-like material with occasional benign epithelial cells, which we interpreted as DPAM. He was then referred to and seen at our centre. He was listed for surgery in a month. Our usual proximal waiting time for patients with DPAM is 3 months but he was classified as urgent because of the obstructive episode.

Three weeks later, he represented to our hospital with current symptoms. He did not have other comorbidities and was not on any regular medications. He was allergic to penicillin, from which he usually develops a rash. There was family history of lung cancer on his maternal side. No other significant family history. He was independent with ADLs and lives at home with wife. He was not a smoker, drinker or drug user. He was still working as a software engineer. No financial issues were identified.

Examination at admission showed a soft and non-tender abdomen, dual heart sound without murmurs. Chest was clear. On the following day, he became febrile with a temperature of 38.3 °C and tachycardia with a rate of 215 bpm (beats per minute) suddenly. The blood pressure was 108/65 mmHg with oxygen saturation of 96% and respiratory rate of 16 bpm. He also complained about diarrhoea and intense pain associated with light-headedness and nausea. Electrocardiogram showed atrial fibrillation with a rapid ventricular rate. His blood results showed leukocytosis, raised CRP (278 mg/L) and decreased haemoglobin (113 g/L). An urgent CT scan and angiogram was ordered and showed multiple new gas pockets in the irregular collection identified previously at the time of diagnosis (Fig. 1). He was suspected to have intraperitoneal sepsis due to infected PMP secondary to rupture of the appendix.

At emergency laparotomy, we found gross intraperitoneal sepsis and did extensive debulking of tumour, appendectomy and extensive division of adhesions (Fig. 2). Another laparotomy was done 24 h later because of his gross sepsis for washout and for the collections in the left and right upper abdomen. Microbiology of surgical swabs showed growth of Enterococcus faecalis, Bacteroides fragilis and Klebsiella pneumoniae. He was subsequently treated with vancomycin, metronidazole and ceftriaxone. Histopathological results of specimens removed at the emergency surgery showed that sections showed dense fibrous connective tissue containing numerous pools or extracellular mucin, many of which contained peripheral strips of atypical mucinous epithelium with neoplastic features.

During ward recovery, his white blood cell count was persistently high. Another CT abdomen and pelvis was ordered and showed small abdominal collections a week after his surgeries. CT-guided drainage was performed. His WCC and CRP counts progressively improved afterwards. He was discharged with oral antibiotics three weeks after his original surgery. He was booked in for outpatient follow-up in four weeks and will be considered for elective cytoreductive surgery (CRS) and perioperative intraperitoneal chemotherapy (PIC) in three months.

Timeline of events was included in Fig. 3.

3. Discussion

Although we have done 780 peritonectomy procedures, this was the first patient we have seen with this presentation of widespread
intraperitoneal sepsis. We decided not to perform a complete cytoreduction in this patient at the time of his presentation with intra-abdominal sepsis and plan a formal CRS and hyperthermic intraperitoneal chemotherapy (HIPEC) at 3 months. We felt that the potential to exacerbate his sepsis made a staged approach likely to be safer. The finding of intermediate grade pathology in this patient was an additional concern, given that true prognosis of such patients is substantially closer to patients with appendiceal adenocarcinoma [4,5].

Intraperitoneal sepsis associated with DPAM, although rare, has been reported [3]. Appendiceal mucinous adenoma develops from appendiceal epithelium and shows progressive growth within the lumen eventually, leading to occlusion of the lumen. Progressive growth of adenoma and continuous production of mucin result in a significant increase in the intraluminal pressure of the appendix, leading to appendiceal rupture and subsequent leakage of mucus into peritoneal cavity. Such an event is usually subclinical. It was suggested that the appendix may repeatedly decompress by perforation and resealing. An episode of appendiceal rupture can cause bacterial contamination of mucus, leading to pan-peritonitis-like symptoms and even, although rarely, sepsis [3,6]. Other more common presentations of PMP include slowly increasing abdominal girth, new ovarian masses and a new onset of umbilical or inguinal hernia [6].

Current standard treatment for PMP is CRS combined with PIC. PIC consists of hyperthermic intraperitoneal chemotherapy (HIPEC) and early postoperative intraperitoneal chemotherapy (EPIC). CRS aims to completely remove all macroscopic peritoneal tumour followed by chemotherapeutic drugs distributed equally inside peritoneal cavity so that microscopic disease can be eradicated [7,8]. Hyperthermia has synergistic anti-tumour effect when combined with certain chemotherapy drugs [8]. EPIC is delivered by some units following CRS and HIPEC, aiming to facilitate further intraperitoneal targeted therapy before the formation of

![Infected PMP (left) and appendix removed (right).](image)

**Fig. 2.** Infected PMP (left) and appendix removed (right).

**Fig. 3.** Timeline of events and management.
postoperative fibrous adhesions [5]. Previous studies have shown that combined therapy has a 5-year survival rate ranging from 57% to 94% [9,10], whereas a recent meta-analysis has shown the mean 5-year survival of this therapy was 76.63% [11].

4. Conclusion

Although bacterial contamination of PMP secondary to appendiceal rupture is rare, we still want to emphasise that surgeons may face an emergency of intraperitoneal sepsis during waiting period of planned CRS or as a primary presentation. With combined therapy of CRS and PIC, the prognosis of mucinous appendiceal adenoma is excellent.

Conflict of interest

None.

Funding

None.

Ethical approval statement

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

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