

Benign multicystic peritoneal mesothelioma associated with hydronephrosis and colovesical fistula formation: report of a case

James C McCaffrey¹, Fung J Foo¹, Neha Dalal², and Kamran H Siddiqui¹

¹Department of General Surgery, ²Department of Histopathology, Tameside General Hospital, Manchester, UK

ABSTRACT

Mesotheliomas usually arise from the pleura and are malignant. We report an unusual case of benign peritoneal mesothelioma presenting in a 59-year-old woman. The disease resulted in bilateral hydronephrosis, colovesical fistula formation, recurrent small bowel obstruction and chronic abdominal pain. To date only a handful of cases have been reported and to the best of our knowledge, none has been so aggressive.

Introduction

Benign multicystic peritoneal mesothelioma (BMPM) is a rare type of tumour arising from the peritoneal mesothelium and was first described in 1979 by Mennemeyer and Smith¹. It usually occurs in young to middle-aged women with a history of pelvic surgery², endometriosis^{3,4} or pelvic inflammatory disease^{5,6}. The aetiology of the condition is controversial, with ongoing discussion as to whether the disease process is primarily reactive or neoplastic.

Although BMPM is associated with a favourable short-term prognosis, and was considered to be a completely benign condition, longer-term follow-up has shown high recurrence rates after surgery^{6,7}. There is also mounting evidence that malignant transformation can occur⁸. No consensus on treatment strategy has yet been reached.

The disease typically follows a slow, progressive course and usually results in chronic abdominal pain. To the best of our knowledge, this is the first reported case of BMPM with such an aggressive nature resulting in bilateral hydronephrosis, colovesical fistula formation, and recurrent small bowel obstruction.

Case report

A 59-year-old woman was admitted with worsening left iliac fossa pain. She had a 6-month history of generalized abdominal discomfort associated with episodes of diarrhoea and 30 kilograms of weight loss. There was no history of prior abdominal or gynaecological surgery, pelvic inflammatory disease, endometriosis or asbestos exposure. Clinical examination revealed a left iliac fossa mass.

CT scan showed multiloculated fluid collection with gas between the bowel loops extending from the lower abdomen into the pelvic cavity posteriorly (Figure 1). High-density contrast in the margins of bowel loops suggestive of diverticular disease was identified. Bilateral hydronephrosis was also noted.

The abdominal mass was initially thought to be a diverticular abscess and the patient was managed conservatively with intravenous antibiotics. When her condition failed to improve, an exploratory laparotomy was performed. This revealed a large cystic mass in the pelvis which had encased the sigmoid colon and rectum. The mass was fixed to the pelvic walls and was unresectable. The uterus and ovaries could not be visualised and ovarian malignancy was suspected. A defunctioning loop colostomy

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Correspondence to: Kamran H Siddiqui, Tameside General Hospital, Fountain Street, Ashton-under-Lyne OL6 9RW, United Kingdom.
Tel +44-161-331 6802;
fax +44-161-331 6610;
e-mail kamran.siddiqui@tgh.nhs.uk

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Figure 1 - CT scan showing multiloculated fluid collection.

my was performed and multiple pelvic biopsy and cytology samples were obtained.

Histology on the endometrial and cervical tissue showed no evidence of neoplasia. Biopsy from the pelvic tumour itself confirmed benign cystic mesothelioma of the peritoneum (also referred to as peritoneal inclusion cysts) (Figure 2). A similar mesothelial inclusion cyst was identified within the omentum. The bilateral hydronephrosis identified was managed by means of ureteric stenting.

Six years after the diagnosis of BMPM, the woman represented with small bowel obstruction. Initial conservative management failed and a laparotomy with adhesiolysis was performed. The intraoperative findings showed aggressive BMPM encasing the pelvic organs.

The patient also began experiencing signs and symptoms of a colovesical fistula, which was confirmed by cystoscopy. A decision against definitive surgical intervention was made, given the findings from the previous laparotomy.

She continued to suffer recurring episodes of small bowel obstruction, which have so far responded to conservative treatment. Urinary tract infections associated with her colovesical fistula have been controlled with low-dose prophylactic oral antibiotics.

Discussion

Mesotheliomas are tumours arising from the epithelial and mesenchymal elements of the mesothelium. The majority of mesotheliomas arise in the pleura, but rare cases of peritoneal², pericardial⁹, and tunica vaginalis¹⁰ mesotheliomas have also been reported. The ratio of the occurrence of peritoneal to pleural tumour is 1:9¹¹. Most mesotheliomas are malignant. Hence, a benign mesothelioma arising from the peritoneum is an extremely rare clinical entity.

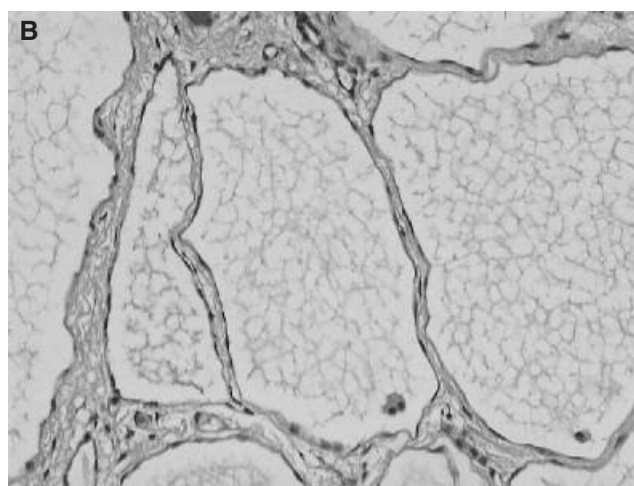
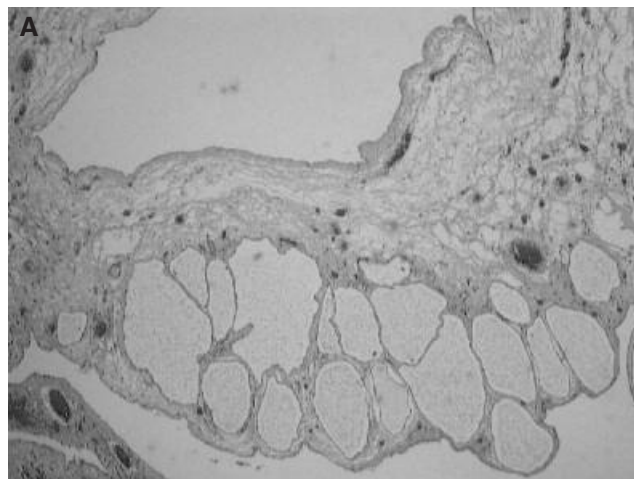


Figure 2 - A) Histological findings by H&E stain (x2.5). Multiple thin-walled locules set in a loose myxoid stroma. B) The locules are lined by a single layer of flattened cells with bland nuclear features. The cells were positive immunohistochemically with calretinin and EMA, confirming their mesothelial origin. H&E stain (x20).

BMPM usually arises in young to middle-aged women (mean 37 years)², although 17% of cases are reported in men¹². Typical presentation is with chronic lower abdominal pain and evidence of a pelvic mass. A history of pelvic surgery², endometriosis^{3,4} or pelvic inflammatory disease^{5,6} is common. BMPM has also been reported to cause partial small bowel obstruction¹³ and acute abdomen⁷.

Disagreement regarding the aetiology of BMPM persists. One hypothesis, supported by the association with previous surgical intervention or chronic infection, suggests a reactive inflammatory aetiology. The other hypothesis suggests BMPM is a neoplastic process. Most current evidence supports the latter hypothesis, with reports of malignant transformation of BMPM⁸, and high post-surgical recurrence rates.

Treatment of BMPM is another area of controversy. Currently, palliative surgical debulking with reoperations for recurrence is the mainstay of treatment. However, some have advocated sclerosive therapy with tetracycline¹⁴, continuous hyperthermic peritoneal perfusion with cisplatin¹⁵, and antioestrogenic drugs¹⁶.

Surgical debulking alone is unsatisfactory for several reasons: i) recurrence rates of 50% are reported^{6,7}; ii) malignant transformation of the lesion can occur⁸ and; iii) patients continue to suffer chronic abdominal and pelvic pain, which tends to be poorly controlled with analgesics¹⁷.

Sethna *et al.*¹⁷ advocate a more aggressive approach aiming for complete disease eradication using cytoreductive surgery with peritonectomy procedures and heated intraoperative intraperitoneal chemotherapy. They describe 5 patients with BMPM who had undergone multiple laparotomies prior to the definitive cytoreduction. All the patients were asymptomatic and 4 out of 5 were disease free with variable follow-up post-cytoreduction ranging from 6 to 69 months.

A review of cases shows that BMPM typically progresses slowly and is not locally invasive. To the best of our knowledge, this is the first case report of BMPM associated with hydronephrosis and colovesicular fistula formation. It highlights the current difficulties faced in managing patients with this condition and urges caution in the assessment of BMPM as a benign process.

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