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Excision of benign multicystic peritoneal mesothelioma and deep infiltrating endometriosis with bowel involvement – A case report

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ABSTRACT

Benign multicystic peritoneal mesothelioma (BMPM) is a rare peritoneal tumour. Treatment involves complete surgical resection, although recurrence rates are high. Notably, there are 7 documented cases of BMPM coexisting with endometriosis on histology and in the case reported here it was associated with deep infiltrating endometriosis. Examination of the 26-year-old nulliparous woman with deep dyspareunia, dyschezia and occasional rectal bleeding revealed restricted uterine mobility and a rectovaginal nodule.

Magnetic resonance imaging (MRI) showed multiple clear cystic structures in the pelvis of unknown aetiology. Following discussion by a multidisciplinary team, a diagnostic laparoscopy was performed. Intraoperatively, bilateral endometriomas were identified, along with multiple fluid-filled cystic structures in the pelvis and on the anterior abdominal wall. An adhesiolysis and drainage of endometriomas was performed and the cystic structures were sent for histology.

Histopathology confirmed BMPM, positive for AE1/3 and calretinin. The patient was referred to a mesothelioma malignancy institute and advised to undergo definitive surgery by the local endometriosis team. A joint surgical procedure with a colorectal team involved laparoscopic excision of peritoneal cysts, cystectomy for bilateral endometriomas, and excision of deep infiltrating endometriosis with bowel shaving. Histopathology revealed benign mesothelial cysts with foci of endometriosis.

The patient had an uncomplicated recovery and is planned for long-term follow-up with the mesothelioma malignancy institute due to the high recurrence rate (up to 50 %). This case report suggests a definitive role of this two-stage procedure in patients with this diagnostic complexity and emphasises the role of multidisciplinary management.

1. Introduction

Cystic mesothelioma is a rare pathology originating from serosal surfaces of the organs such as peritoneum, pleura and pericardium. The commonest variety is peritoneal mesothelioma, which disproportionately affects women (5:1). It is usually seen in women of reproductive age, given that the average age at diagnosis is 32 [1].

Benign multicystic peritoneal mesothelioma (BMPM) comprises 3–5 % of these peritoneal mesotheliomas. Although the exact aetiology is not known, an association has been found with endometriosis, pelvic inflammatory disease and previous surgeries [2]. In the majority of these cases, imaging is inconclusive, and the diagnosis is confirmed histologically [3,4]. Histology shows multiple cysts lined by bland mesothelial cells and expression of pankeratin Cam 5.2 and mesothelial cell markers, calretinin and WT-1. [5].

It is a very rare disease, with an annual incidence of 1.2 per million [6]. The rarity and diversity of these lesions along with variable behaviour serves as a diagnostic dilemma and warrants accurate diagnosis. Therefore, it is very important that these cases are optimally reported and discussed in the literature. We are reporting the what seems to be the first case associated with deep infiltrating bowel endometriosis and the seventh case with histologically proven endometriosis.

2. Case Presentation

A 26-year-old woman presented with severe lower abdominal pain, dysmenorrhea, deep dyspareunia, dyschezia and occasional rectal bleeding. Examination showed restricted uterine mobility and a

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rectovaginal nodule. A Mirena coil was placed to help with her cyclical symptoms, but symptoms persisted. She was started on gonadotrophin-releasing hormone analogues (GNRHa) with add-back hormone replacement therapy (HRT), and was given a preliminary diagnosis of endometriosis.

Magnetic resonance imaging (MRI) revealed multiple clear cystic structures in the pelvis, adenomyosis, bilateral endometriomas and adhesions between the uterus and posterior rectosigmoid junction (Figs. 1 and 2). Diagnostic laparoscopy was performed as imaging was not entirely consistent with the diagnosis of endometriosis.

Primary laparoscopy revealed haemosiderin deposits throughout the pelvis and peritoneum of the upper abdomen and diaphragm. There was a large, left endometrioma, approximately 10 cm, with the sigmoid adherent beneath the left endometrioma. The right endometrioma, approximately 6 cm, was overlying the fundus of the uterus (Fig. 3). With mobilisation of the endometriomas, it was evident that the rectum was adherent to the uterus. The pouch of Douglas was almost completely obliterated except for a small window of access to the left of the rectum. There appeared to be thickening over both uterosacral ligaments, both being tethered towards the uterus. On rectal examination, a plaque could be felt anteriorly at about 10 cm. There were limited views at rigid sigmoidoscopy.

Both endometriomas were freed and drained. There were fluid-containing peritoneal cysts throughout the pelvis, one on the anterior abdominal wall, two small ones within the pouch of Douglas, and some para-fimbrial cysts on the right. These were all removed and sent for histology.

Postoperatively, the patient continued on GnRHa with add-back HRT, and follow-up was organised. The histology report showed benign peritoneal multicystic mesothelioma positive for AE1/3 and calretinin. The patient was referred to a national mesothelioma Centre and discussed in the national peritoneal mesothelioma multidisciplinary team (MDT). Options for treatment included conservative management or a further surgery to remove any peritoneal disease. However, as the patient remained symptomatic due to untreated endometriosis, a second surgery was planned as a joint gynaecological and colorectal procedure. Interim flexible sigmoidoscopy did not reveal any mucosal involvement

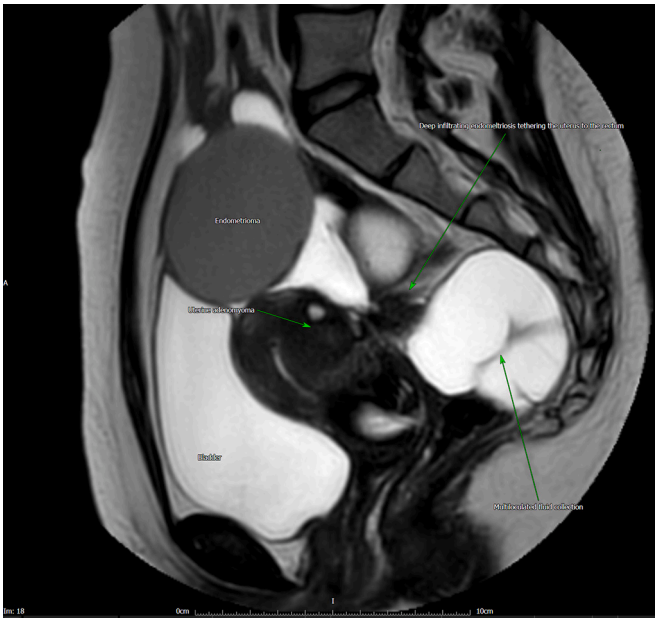

Fig. 1. T2 sagittal magnetic resonance image through the mid-pelvis. The uterus has a large focal area of adenomyosis and is tethered to the rectum by deep infiltrating endometriosis. Posterior to this, there is a multiloculated fluid collection, histologically proven mesothelioma. A separate endometrioma is labelled cranial to the bladder and uterine fundus.

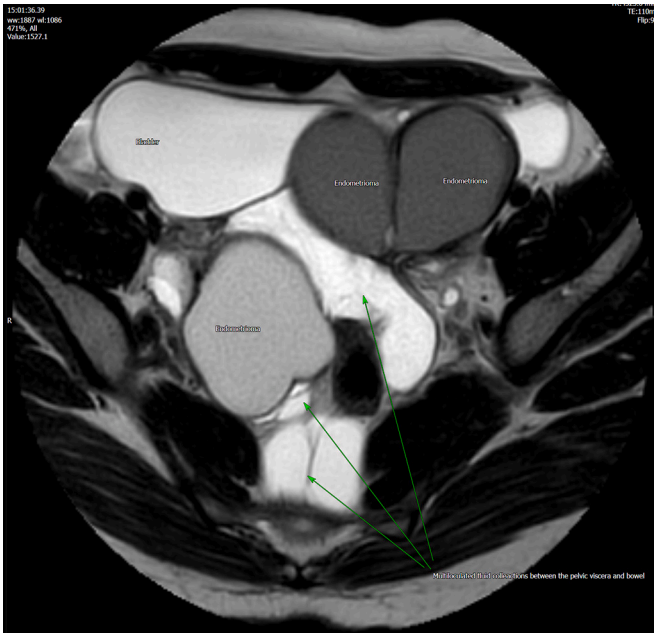

Fig. 2. T2 axial magnetic resonance image through the mid-pelvis shows bilateral endometriomas with intervening multiloculated fluid collections, histologically proven to represent mesothelioma.

of the rectum.

At second laparoscopy, both endometriomas were found to have recurred (Fig. 4) and were adherent to the posterior uterus, sigmoid and rectum. There were further multiple small peritoneal cysts close to the fimbrial end of the fallopian tubes and there was a small area of endometriosis in the right UV fold. The rectum was completely adherent to the uterus, obliterating the pouch of Douglas. Both ovaries were carefully mobilised with drainage of bilateral endometriomas. All visible pelvic cysts were excised. Both ovaries were suspended, the sigmoid was mobilised and bilateral ureterolysis was performed. The pararectal space was dissected bilaterally and the bowel was freed from the back of the uterus. The endometriotic nodule was shaved from the wall of the rectovaginal septum. The serosa was reinforced with 3 interrupted PDS sutures. A rectal air test was performed and showed rectal wall integrity. All remaining endometriosis was dissected from the pelvic side wall, both uterosacral ligaments and back of the uterus. The ovaries were released and the endometriomas were bluntly stripped with careful diathermy to the base (Fig. 5).

The patient made an uneventful postoperative recovery. She will be under long-term follow-up at the national mesothelioma specialist centre and local endometriosis centre.

3. Discussion

BMPM is a rare entity discussed by Mennemeyer et al. in 1979 for the first time [7]. Only 200 cases have been reported to date, with majority being female. The commonest presenting features has been vague lower abdominal pain [8]. Other features include fullness, distension, pelvic pain, palpable mass, weight loss, nausea, vomiting and constipation [8–13]. Reports of cases presenting with bowel obstruction, urinary retention and acute abdomen have been published [14–17]. The patient in the present case presented with long-standing pelvic pain but because of endometriosis there was significant overlap of symptoms.

The aetiology of benign mesothelioma is not clearly understood and there is debate about its reactive or neoplastic nature. As it is often seen in women with prior pelvic pathologies and surgeries there is greater consensus about its reactive nature [7,8]. The malignant forms are seen associated with asbestos exposure [18].

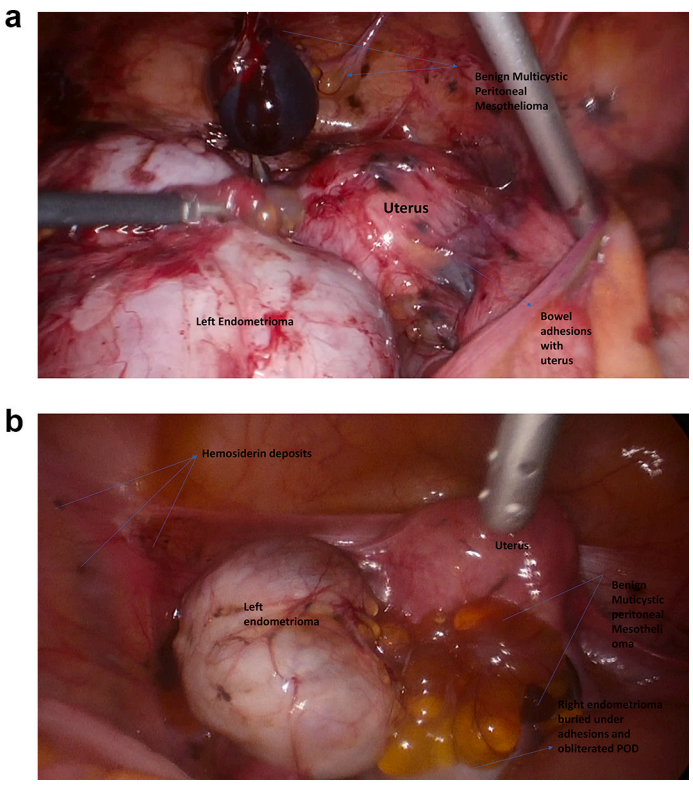

Fig. 3. (a and b) Primary laparoscopy.

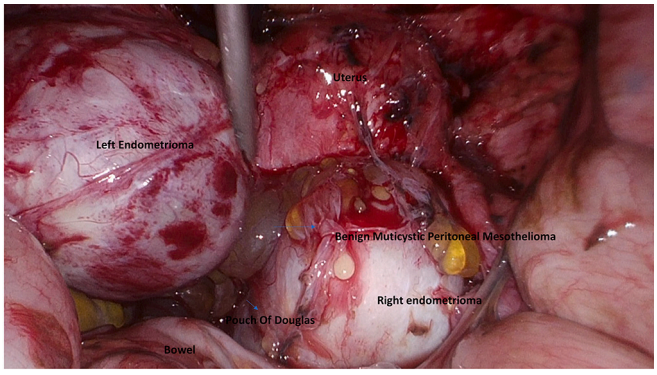

Fig. 4. Secondary laparoscopy.

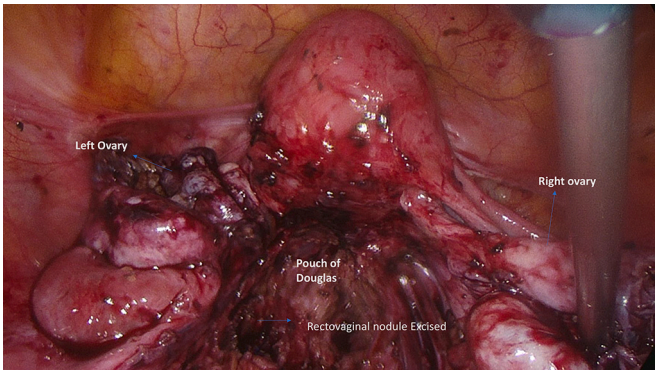

Fig. 5. Image from the end of the procedure.

Although ultrasound, computed tomography (CT), and MRI demonstrate an abnormality suggestive of this disorder, confirmation of the diagnosis is accomplished intraoperatively [19]. In this case, MRI showed multiple clear cysts of unclear cause. The diagnosis was established on histology, which is consistent with the current literature, where in majority of the cases the disease was identified intraoperatively or postoperatively.

Management of the disease varies from conservative management, hormonal therapy such as antioestrogen or gonadotrophin to complete resection/cytoreductive surgery and hyperthermic intraoperative intraperitoneal chemotherapy (HIPIC). Although the disease is known to have a high recurrence rate [20], there is paucity of evidence as to prognosis, optimal management and follow-up. In this patient, very few peritoneal cysts were present at the follow-up surgery, which suggests that removing the primary pathology such as endometriosis may result in natural resolution of the disease and aggressive therapy should be considered with caution [7,20], especially in view of low malignant transformation.

Despite the quality of pre-operative diagnostics in endometriosis and the trend towards 'see and treat' laparoscopy, it is appreciated in cases such as the present one that there is a definite role for a diagnostic procedure, where imaging was not conclusive of diagnosis. This meant that biopsies could be obtained for histological diagnosis and a secondary definitive surgery could be safely planned to optimise outcome.

This case is reported to highlight the difficulty surrounding diagnosis and management. This is apparently the first reported case of BMPM with deep infiltrating endometriosis involving the bowels.

4. Conclusion

BMPM is a rare condition associated with endometriosis. MDT involvement is crucial in managing complex and rare cases such as BMPM with co-existing endometriosis. There is also a definitive role for a two-stage procedure in cases with diagnostic difficulty. Endometriosis centres should try to establish a database for these rare disorders and

develop consensus management and follow-up plans.

Contributors

Zahra Azeem contributed to patient care, conception of the case report, drafting the manuscript, undertaking the literature review and revising the article critically for important intellectual content.

Jyoti Sharma contributed to patient care, undertaking the literature review and revising the article critically for important intellectual content.

Rob Johnson reported the MRI images and provided images for publication.

Natalia Price contributed to patient care,

Miquel Zilvetti Yabar contributed to patient care.

Donna Ghosh contributed to patient care, conception of the case report, and revising the article critically for important intellectual content.

All authors approved the final submitted manuscript.

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Patient consent

Obtained.

Provenance and peer review

This article was not commissioned and was peer reviewed.

Conflict of interest statement

The authors declare that they have no conflict of interest regarding the publication of this case report.

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