CASE REPORT

The curious case of free-floating pelvic cysts

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Accepted 30 May 2014

SUMMARY

The case of a healthy 33-year-old woman who underwent an elective caesarean section with incidental finding of small free-floating cysts within the pelvic peritoneum is presented. Gross examination could not identify the specimens. Histopathological investigation was necessary to classify the specimens as benign mesothelial cysts. The patient had no medical or surgical history and no other risk factors. The presented case is particularly interesting as the cysts were non-adherent to any pelvic or abdominal structure, as is commonly described. Similarly, there are few descriptions of mesothelial cyst diagnosis during pregnancy.

BACKGROUND

Benign multicystic peritoneal mesothelioma (BMPM) is a rare tumour. To date, it has mostly been described in women of reproductive age. It may be found incidentally on imaging or intraoperatively in an otherwise asymptomatic patient. Equally, it may present with an abdominal mass or pain, necessitating surgery as the first-line management option. Imaging cannot distinguish BMPM from other cystic structures and pathological examination is required for a conclusive diagnosis. The cysts may be recurrent in nature even after complete surgical removal. There is controversy as to the pathogenesis of BMPM in whether the cysts are part of an inflammatory process affecting peritoneal cells, or whether they should be classified as a neoplastic process.

Our presented case adds to the literature that mesothelial cysts are benign, often asymptomatic and most commonly found in women of reproductive age. It also suggests that these cysts may appear unattached to pelvic or abdominal structures. Importantly, the literature describes few cases of mesothelial cyst discovery in pregnancy.

CASE PRESENTATION

A healthy 33-year-old Caucasian woman underwent an elective caesarean section at term plus 6 days gestation. The indication for the caesarean section was a failed induction of labour, which had been started due to worsening liver function and pruritus suggestive of cholestasis of pregnancy. The patient had no significant medical or surgical history, no regular medications and no known drug allergies. She was a non-smoker with no alcohol intake during pregnancy. A lower segment caesarean section was successfully performed under spinal anaesthesia delivering a healthy baby girl weighing 3520 g. There were no obstetric or anaesthetic complications. There was an estimated blood loss of 750 mL. After closing the uterotomy the

ovaries and fallopian tubes were visually examined as per regular practice, all anatomy appeared normal. During the examination, four small cystic structures were found apparently free floating within the pelvis. The four cysts were found in two pairs with each pair being joined by a flimsy band of tissue. They were non-adherent to any pelvic or abdominal structure and appeared intact with no evidence of trauma. Grossly, the cysts were thin walled with a defined ovoid shape filled with a clear serous fluid and appeared unilocular in nature. No other abnormalities were noted in the abdomen. The cystic structures were sent to histopathology for classification. The patient described no abdominal or pelvic symptoms out with the normal expected during pregnancy and no symptoms prior to conception.

INVESTIGATIONS

All specimens were sent to histopathology for further investigation. Grossly (figure 1), the cystic structures were collapsed measuring 9×4×0.5 and 21×2×1 cm. The specimens were partly filled with clear fluid. The cyst walls had occasional areas of firmer grey tissue but no solid areas were identified. Representative tissue samples were isolated and fixed in formalin, then embedded in paraffin prior to staining with H&E. Microscopically, the structures were composed of fibroconnective tissues (figure 2) with multicellular lining. The wall was lined by regular flattened to cuboidal cells that showed no atypia and exhibited no mitotic features (figure 3). On immunohistochemical staining there was expression of calretinin, which is a selective mesothelial marker (figure 4). The specimens lacked smooth muscle. Importantly, there were no features of malignancy in any of the tissue samples.

DIFFERENTIAL DIAGNOSIS**Benign**

- ▶ Cystic lymphangioma
- ▶ Cystic adenomatoid tumours
- ▶ Cystic endosalpingiosis
- ▶ Endometriosis
- ▶ Mullerian cysts
- ▶ Pseudomyxoma peritonei
- ▶ Cystic mesonephric duct remnants.¹⁻³

Malignant

- ▶ Mesothelioma
- ▶ Serosus tumours of the peritoneum
- ▶ Ovarian clear cell carcinoma.^{1 2}

The above differentials are distinguished by clinical history and importantly histopathological investigation. The most prudent conditions to exclude are cystic lymphangiomas and cystic adenomatoid tumours. As discussed, BMPM is a benign cyst

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To cite: Watson HI, Borovickova M, Shetty A. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2014-205229

Figure 1 The specimens are seen in a pathology sample container. Grossly, the specimens were partly filled with clear fluid.

characterised by cuboidal mesothelial cells with a distinctive lack of smooth muscle. In contrast cystic lymphangiomas, which represent malformations of lymphatic vessels, contain bundles of smooth muscle. These will be positive for CD34 emphasising the endothelial derivation of this benign tumour. Comparatively, adenomatoid tumours rarely present with a cuboidal epithelial lining. They are more glandular in origin and so more commonly contain pseudoglandular spaces containing basophilic secretions. Additionally, they may be interspersing with connective tissues and hyperplastic smooth muscle cells. The other benign differentials are histologically differentiated from BMPM by identification of the cell origin of each pathological diagnosis.⁴

It is clear that the most important distinction remains between BMPM and its malignant mimickers. Histologically, the malignant differential diagnoses will display typical malignant features such as cellular pleomorphism, nuclear atypia and high mitotic activity; all these features are absent from BMPM.⁴

TREATMENT

Given the benign nature and asymptomatic presentation of these cysts no further management was required for this patient.

Figure 2 Histopathology slide at $\times 2$ magnification. The cyst wall is thin and is composed of fibroconnective tissues.

Figure 3 Histopathology slide at $\times 20$ magnification. The wall of the cyst is lined by regular flattened to cuboidal mesothelial cells. Importantly, the cells show no atypia or mitotic features.

OUTCOME AND FOLLOW-UP

The patient has been educated on the possible recurrence of the cysts and potential presentation symptoms. The specimens showed no evidence of malignancy and no specific follow-up was merited. Similarly, no additional obstetric follow-up was required postoperatively. The patient was seen routinely by her community midwife with no postnatal issues.

DISCUSSION

The majority of the evidence regarding BMPM is derived from case reports and short case series. Importantly, BMPM has been given multiple names since it was first described in 1979 making interpretation of the literature ambiguous at times. In 1979, Mennemeyer⁵ first described these lesions as 'multicystic peritoneal mesothelioma'. Throughout this report, BMPM will be used to collectively discuss this topic.

BMPM is a rare condition but of the reported cases the majority are in women of reproductive age, with few cases in men and postmenopausal women. Owing to this pattern it has been suggested that the cysts are at least in part hormonally driven, likely by oestrogen. Given this, it is surprising that there are very few reports of mesothelial cyst occurrence during pregnancy.⁶ One report described the incidental finding of a mesothelial cyst in the right iliac fossa during a caesarean section.

Figure 4 On immunohistochemical staining the tissue sample expressed calretinin: a selective mesothelial marker.

This patient differed from the reported patient as the cyst was adherent and the woman had a surgical history of hysterosalpingography and hysteroscopy.⁶

Although there is controversy the overall suggestion is that these cysts are benign in nature; this will be discussed further. There are numerous case reports of these cysts recurring despite successful operative removal with some patients requiring multiple operations to reduce the cystic load and relieve symptoms. Our patient did not follow this pattern given her asymptomatic incidental presentation. Similarly, many reports describe a pelvic or abdominal mass being the presentation symptom and this is partly because the lesion may be adherent to an organ or omentum and so stabilised in one location. Again, the described patient differs in that the lesions were found as free-floating entities within the peritoneum. Mesothelial cysts have been found out with the pelvis, namely, pleural and attached to the pericardium. They are named according to their anatomical location, such as, the pericardial cyst.

Some specialists suggest that there are at least two distinct types of multiple cysts that are associated with chronic pelvic pain. The first is recognised in patients who have undergone multiple pelvic surgeries and whose cysts are deemed to be partly causal to adhesions postoperatively. These have been given the names of postoperative peritoneal cyst or peritoneal inclusion cyst. The other form described may be more in keeping with our patient, who had not undergone any operative intervention. These are termed benign cystic mesothelioma or multicystic peritoneal mesothelioma and are thought to be related to inflammatory or reactive processes.⁷

Of the benign differentials, cystic lymphangioma and cystic adenomatoid tumours are considered the most important to exclude. In fact, the first description of BMPM was related to a case report from 1979 detailing how BMPM could masquerade as a cystic lymphangioma. At this time, electron microscopy was used to differentiate between the two cystic lesions.⁵ The two are distinguished largely by anatomical location, in that cystic lymphangiomas are rarely found in relation to the pelvis. As discussed above, microscopically areas of smooth muscle and lymphoid tissue can be identified. Cystic adenomatoid tumours may be commonly confused with mesothelial cysts, on macroscopic and microscopic levels. However, adenomatoid tumours usually contain a solid component, unlike BMPM.¹

Importantly, it has been suggested in the literature that BMPM may be placed on a spectrum between an adenomatoid tumour and a malignant mesothelioma.⁸ To date, there have been only a few descriptions of malignant transformation of BMPM. One involved a 36-year-old woman who underwent six surgical procedures to lessen the cystic load with eventual transformation into a malignant mesothelioma with aggressive, diffuse involvement of previous abdominal wound incisions and lymph nodes.⁹ This report did not mention any exposures to carcinogens, such as asbestos that might be related to the malignant transformation.

Interestingly, the debate surrounding the pathogenesis of BMPM was evident in the first reported case back in 1979. The authors thoroughly ruled out a reactive cause for the mesothelial cell proliferation and so deemed the lesions to be a neoplastic process.⁵

Since the first description of BMPM there has been suggestion that a strong hormonal influence plays a role in their formation and perpetuation. A case report from 1995 described a 17-year-old girl with recurrent peritoneal mesothelial cysts. The cystic load could be reduced with the use of gonadotrophin-releasing hormone (GnRH) agonists and the cyst volume would

increase with 'add-back therapy' consisting of oestrogen and progestin. On reintroducing the GnRH agonist cyst size once again reduced.¹⁰ GnRH agonists essentially induce a hypo-oestrogenic state that favours reduction in cyst volume size but negatively affects other physiological parameters, such as bone mineral density. GnRH agonists would only ever be suitable for short-term use and it has been suggested that they could be used preoperatively to reduce cyst volume and aid operative success. However, this modality is likely unsuitable for the majority of patients given the hormonal and other side effects. A more long-term alternative may be a drug with agonist and antagonist properties towards oestrogen, such as, tamoxifen. Its successful use has been reported in one case study.¹¹ While GnRH agonists could only ever be a short-term measure, tamoxifen would be suitable for more prolonged use given the positive preservation of bone and cardiovascular health. Additional evidence on hormonal dependence is also derived from cyst recurrence in menopausal women, suggesting that peripheral as well as central oestrogen may play a part in cyst perpetuation.¹¹

Management of BMPM may therefore range from conservative to medical or surgical treatment options. Conservative management may well be appropriate in patients such as ours that presented incidentally with no concerning history or distressing symptoms. In the more problematic cases, medical options, including GnRH agonists and drugs with antioestrogen activity like tamoxifen, provide compelling evidence as to the pathogenesis of BMPM but concerning side effects must be considered in the risk analysis. Within the literature, the most common management option appears to be surgical removal of cysts.

Learning points

Mesothelial cysts are:

- ▶ Often asymptomatic and discovered incidentally.
- ▶ Benign in nature.
- ▶ More common in women of reproductive age.
- ▶ Uncommonly found to be apparently free floating in the peritoneum.
- ▶ Not always associated with previous surgery or inflammatory conditions.

Contributors All authors equally contributed to the writing, research and editing of this case report.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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