Benign multicystic peritoneal mesothelioma (BMPM) – case report and review of the literature

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Benign multicystic peritoneal mesothelioma (BMPM), also known as multilocular peritoneal inclusion cyst, is a rare tumour that occurs mainly in women at their reproductive age. The aetiology and pathogenesis are controversial. It originates from any abdominal peritoneal or pleural surface. The biological behaviour of BMPM is usually clinically benign. Here we present a case report of BMPM from our department with a review of the literature.

Key words: benign multicystic peritoneal mesothelioma, multilocular peritoneal inclusion cyst, peritoneum, omental cystic masses.

Introduction

Benign multicystic peritoneal mesothelioma (BMPM), also known as multilocular peritoneal inclusion cyst, was first described in 1928 by Plaut, who came across a collection of the thin-walled cysts as an incidental finding during surgery for uterine leiomyomas [1]. It is a rare tumour that occurs mainly in women (83%) at their reproductive age [2]. The aetiology and pathogenesis are controversial regarding its neoplastic and reactive nature. It originates from any abdominal peritoneal or pleural surface [3-5]. The biological behaviour of BMPM is usually clinically benign. However, this lesion is characterized by its slowly progressive nature and high rate of recurrence after surgical resection [6].

Case presentation

A 46-year-old female was referred to the hospital with the diagnosis of a painful mass in her abdomen. The patient underwent a total abdominal hysterectomy, bilateral salpingo-oophorectomy and partial omentectomy. The hysterectomy specimen sent for the histological examination consisted of irregular multilocular thin-walled fluid-filled cystic mass arranged in grape-like clusters attached to the greater omentum. The cysts ranged from 0.2 to 3.5 cm in their greatest dimension (Fig. 1). No solid areas, necrosis or haemorrhage were identified.

The specimen was fixed in a solution of 10% buffered formalin for 3 days. Representative sections were chosen for histopathological examination. Paraffin blocks were prepared and sliced into 4 µm thick sections by a microtome and stained with haematoxylin and eosin. Immunohistochemical stainings were performed on paraffin sections using a DAKO Immunostainer and primary antibodies AE1/AE3 (DAKO), ER (DAKO), PR (DAKO). Histological examination of the lesion revealed multiple cysts covered by flat to cuboidal mesothelium-like epithelium with no malignant features. The cysts varied in size and were filled with amorphous eosinophilic material. The cysts were surrounded by fibrovascular stroma. Endometrium was in the proliferative phase and myometrium showed foci of adenomyosis and leiomyomas. Immunohistochemically mesothelial cells stained positively for cytokeratins (Fig. 2) but did not immunoreact to progesterone and oestrogen (Fig. 3, 4). The pathologic diagnosis was multicystic mesothelioma of the peritoneum.

Discussion

Plaut first described multiple peritoneal cysts in 1928 but their mesothelial nature was confirmed by Menemeyer and Smith in 1979 [1, 7]. BMPM is a rare condition occurring mainly in women at child-bearing age and is associated with a history of previous abdominal surgery, endometriosis, leiomyomas or pelvic

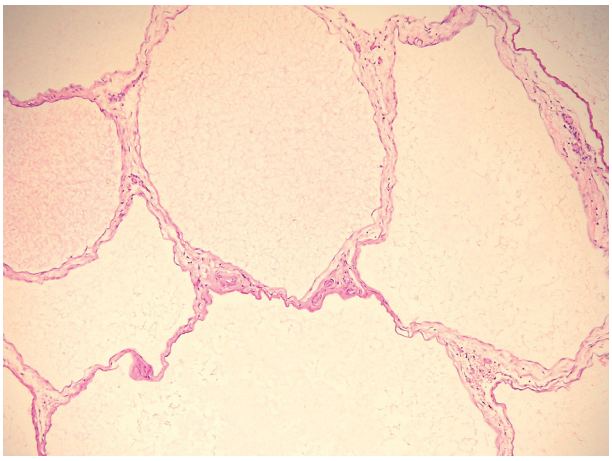

Fig. 1. Variable-sized cysts lined by flattened, mesothelial cells and filled with amorphus eosinophilic material. Haematoxylin and eosin, original magnification 100×

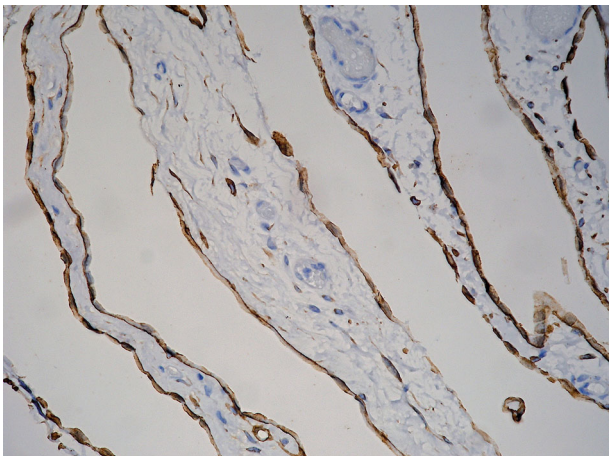

Fig. 2. Immunohistochemical staining shows the presence of cytokeratin in the cytoplasm of the lining cells. AE1/AE3, original magnification 400×

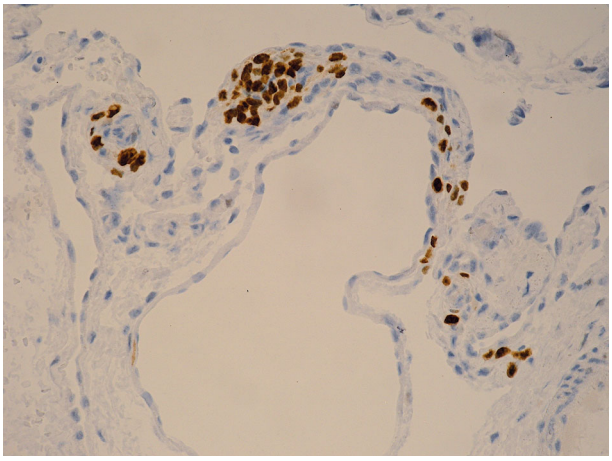

Fig. 3. Progesterone-positive cells in a delicate fibrous stroma. Mesothelial cells are negative for progesterone. PR, original magnification 400×

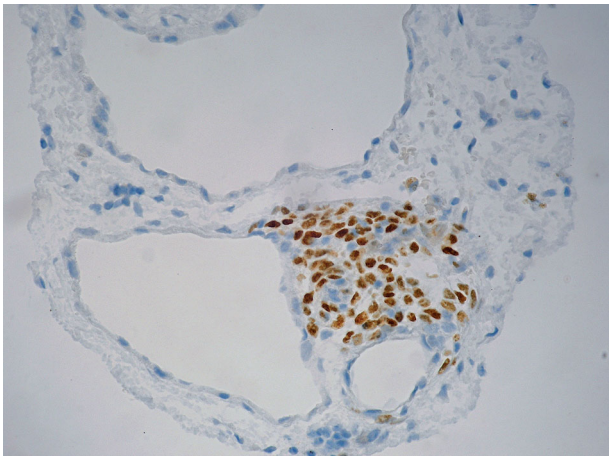

Fig. 4. Oestrogen-positive cells in a delicate fibrous stroma. Mesothelial cells are negative for progesterone. ER, original magnification 400×

inflammatory disease (PID) [3, 8, 9]. However, the lesion has also been described in men, infants and in patients in their 60s. They are usually localized in the pelvic peritoneum but also on the surfaces of the uterus, ovary, bladder, rectum, or cul-de-sac [10]. Sometimes the retroperitoneum is involved. A similar lesion has been described in the pleural cavity. Unlike pleural mesothelioma, this condition is not asbestos related [1].

The aetiology and pathogenesis are controversial regarding its neoplastic and reactive nature [3-5]. The progressive growth, a marked tendency to recur especially after an incomplete excision, a low incidence of abdominal infection and a high disease-related mortality suggest a neoplastic origin [3, 6, 10]. Microscopically revealed inflammation, areas of endometriosis or uterine leiomyomas and a history of previous abdominal surgery support its reactive nature. It is considered to be a peculiar peritoneal reaction to chronic irritation stimuli, with mesothelial cell entrapment, reactive proliferation and cystic formation [3-5].

The differential diagnosis should include such benign and malignant conditions as lymphangioma, other mesenteric-omental cysts, cystic forms of endosalpingiosis, ovarian cystadenoma or cystadenocarcinoma, cystic teratoma, pseudomyxoma peritonei, cystic smooth muscle tumours, visceral cysts, cystic mucinous neoplasm of the pancreas, endometriosis and echinococcal cysts [2-4, 6, 7, 10-15]. In fact, the most important differential diagnosis is between BMPM, cystic lymphangioma, cystic adenomatoid tumour and malignant mesothelioma. Pathologically, lymphangioma is mostly a large, thin-walled, cystic mass, usually multiloculated. On gross examination the cystic component of lymphangioma is predominantly chylous (but may be serous or hemorrhagic) and microscopic examination reveals cystic spaces covered by a single layer of flattened endothelial cells (CD31+, CD34+, Factor VIII+, VEGFR+), bounds of smooth muscle and aggregates of lymphocytes in its wall [2-4, 6, 11, 13]. An adenomatoid tumour is a lesion of mesothe-

lial origin, four types of which have been described: adenoid, angiomatoid, solid and cystic. It is observed predominantly in the male genitalia (epididymis) but can also appear in females (fallopian tubes, uterus, ovary, omentum and mesentery). A cystic variant of this tumour reveals spaces lined with flattened to cuboidal cells but often a recognizable solid component can be also found [3, 4, 6, 11, 12]. The most important lesion in the differential diagnosis is cystic malignant mesothelioma. The median age at diagnosis is 60, and the male : female ratio is 3 : 1. The majority of patients have a history of previous asbestos exposure, although this association is much less common in females. On gross examination, malignant mesothelioma usually reveals numerous small, shiny nodules, but cystic structures with extensive microcystic changes can also be found. Malignant form of mesothelioma can be distinguished from BMPM by the presence of malignant features such as destructive growth (infiltration of the entire omentum), cellular atypia and increased mitotic count with abnormal mitoses [3, 4, 6, 11, 14, 15].

The diagnosis of BMPM is most effectively accomplished through the use of a CT scan. A correct pre-operative diagnosis is almost never rendered but some of these lesions have characteristic radiological features that help to differentiate them from BMPM [6, 10, 11, 16].

BMPM is a benign, probably reactive condition with slowly progressive nature, high rate of recurrence after surgical resection and only one fatal outcome reported in the literature [1, 5]. One case with malignant transformation of BMPM has been reported [17].

Total surgical excision, particularly of the localised disease, is the most effective treatment, but the local recurrence rate is fairly high, between 33 and 50%. Chemotherapy and radiotherapy are not effective in this case [3, 10, 11, 18, 19].

It is important to recognise this condition correctly as it occurs in young women in whom fertility must be taken into consideration in pelvic surgery [20].

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