Turk J Gastroenterol 2009; 20 (2): 138-141

Benign multicystic peritoneal mesothelioma

Peritoneal benign kistik mezotelyoma

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Benign multicystic peritoneal mesothelioma is a rare tumor that occurs mainly in women in their reproductive age. It is characterized by the formation of multiple, thin-walled, multilocular cysts that frequently produce large, intra-abdominal masses. The short follow-ups and possible etiologies based on the published reports make it difficult to draw any firm conclusions.

Key words: Benign multicystic peritoneal mesothelioma, immunohistochemistry, peritoneum

Benign kistik mezotelyoma çoğunlukla üreme çağındaki kadınlarda görülen nadir bir tümördür. Büyük intra-abdominal kitleler yapan; çok sayıda, ince duvarlı multiloküler kistler ile karekterlidir. Yayınlanan vakalardan belli sonuçlar çıkarmak, kısa dönem takip süresi ve olası etyolojilerin varlığı nedeniyle güçtür.

Anahtar kelimeler: Benign kistik mezotelyoma, immünohistokimya, periton

INTRODUCTION

Benign multicystic peritoneal mesothelioma (BMPM), also known as multilocular peritoneal cysts, is an uncommon lesion arising from the peritoneal mesothelium. This lesion occurs most frequently in women in the reproductive period (1). The etiology, pathogenesis and biological behavior of BMPM have remained a great mystery (2). To date, approximately 130 cases have been reported (3). We herein report a case of a 48-year-old woman who presented with abdominal pain and swelling.

CASE REPORT

A 48-year-old woman was admitted to the surgical department due to abdominal pain and abdominal swelling. On admission, she reported no menstruation history for 18 months. She had been having a history of frequent parasitic infections since childhood. She had no medical history of malignancy. There was no discretely palpable mass on physical examination. Laboratory data were within normal ranges. Abdominal ultrasonography demonstrated a cystic-appearing mass arising from the peritoneum. Thickness in the omentum and cystic structures were observed on the computerized tomography (Figure 1). Grape-like cystic structures were seen in laparoscopic examination and incisional biopsy was performed. The differential diagnoses considered at this point were peritonitis and malignancy. Cytopathologic examination of abdominal fluid did not demonstrate any malignant cells. The pathologic diagnosis was consistent with BMPM and thus, total omentectomy operation was performed.

Two materials were obtained from the patient: the first incisional specimen measured 3 x 2.5 cm and 1 cm in diameter, while the second omentectomy material measured 35 x 17 cm and 1 cm in diameter. Gross examination of these specimens revealed a multicystic mass. Individual cysts ranged from 0.3 to 2 cm in diameter and contained serous fluid. The walls of the cysts were semi-transparent and less than 1 mm thick. No solid areas were found within the lesion (Figure 2).

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Manuscript received: 30.11.2007 **Accepted:** 08.05.2008

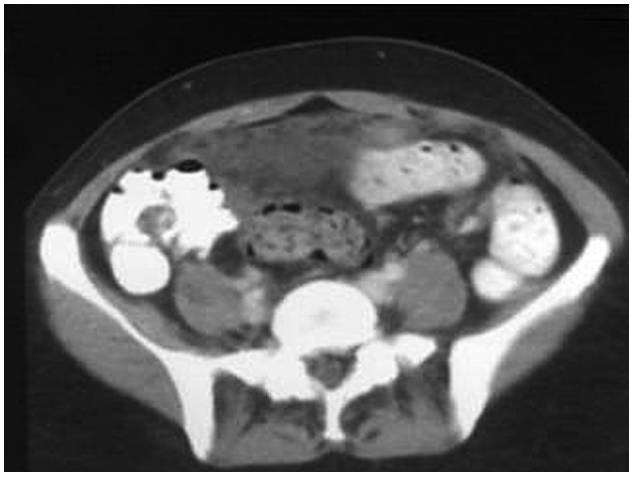

Figure 1. Computerized tomography shows cystic structures and thickness in the omentum.

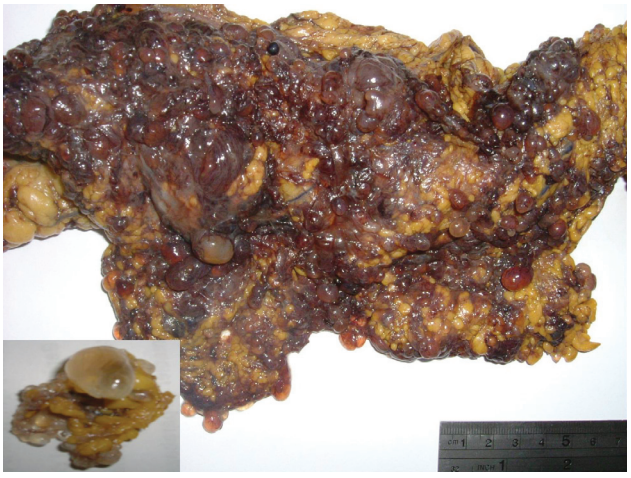

Figure 2. Gross appearance of both incisional (insert) and omentectomy specimens.

The histological appearances of the tumors within the biopsy specimen and omentectomy material were similar. The walls of the cysts were lined by single flat cells alternating with cuboidal cells with hobnail features; no atypia or mitosis was noted (Figure 3). The septa consisted of loose connective tissue. Scattered lymphocytes were observed within this tissue. Vascular congestion was observed (Figure 4). The mesothelial nature of the lining cells was confirmed by positive immunohistochemical staining for cytokeratin (Figure 5) and calretinin. CD34 was negative in these cells.

The patient had an uneventful recovery and there was no recurrence two years after total omentectomy operation. In addition, she has had regular menstruation for the last six months.

DISCUSSION

Benign multicystic peritoneal mesothelioma was first described in 1979 by Mennemeyer and Smith (4). This disease is classified as an exceedingly rare medical entity, which renders its origin, pathogenesis, diagnosis and therapy challenging (3).

These lesions occur predominantly in women in the reproductive age group. The most common presenting complaints are pelvic and low abdominal pain, but the lesions are sometimes incidental findings at laparotomy (1, 5, 6). However, cases involving men, preadolescent children (7) and elderly women (8) and rare extra-abdominal cases have been reported (7).

Most patients are diagnosed incidentally during

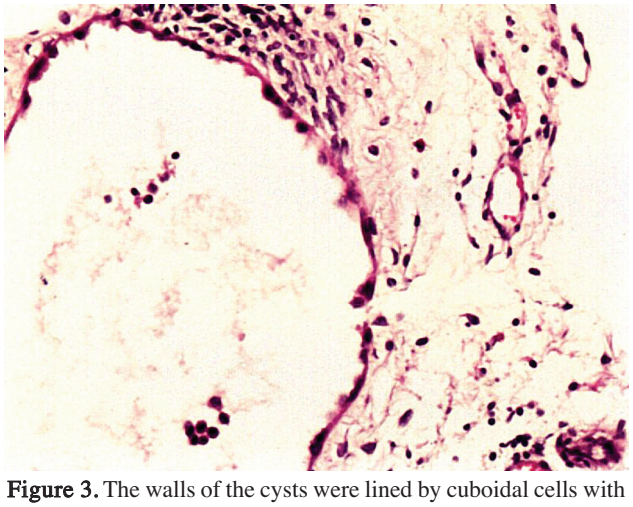

Figure 3. The walls of the cysts were lined by cuboidal cells with hobnail features. Scattered lymphocytes were observed in the stroma (hematoxylin and eosin x400).

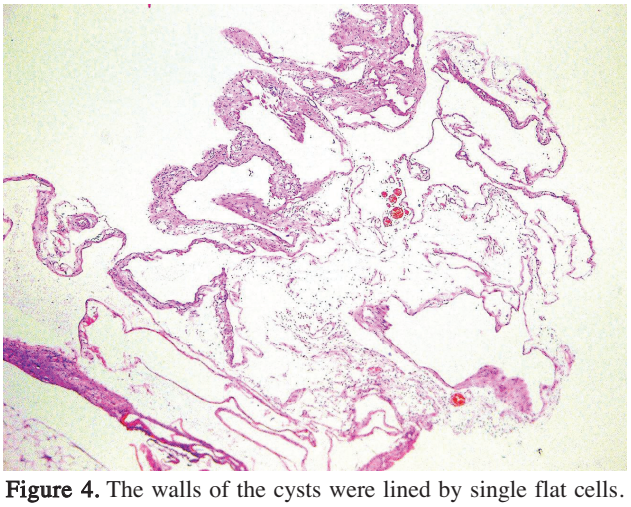

Figure 4. The walls of the cysts were lined by single flat cells. The septa consisted of loose connective tissue (hematoxylin and eosin x40).

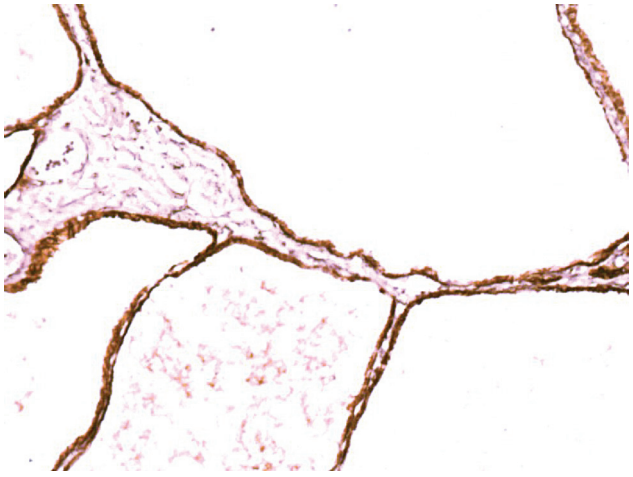

Figure 5. Strong membranous immunoreactivity for cytokeratin in cells lining the cysts (DAB x 200).

examination or laparotomies for other reasons (3). Rarely, weight loss and a huge abdominal mass can prompt the diagnosis (9). A differential diagnosis from other cystic neoplastic or inflammatory lesions is difficult even with modern imaging techniques (3, 10). No relation with asbestosis has been found (11).

BMPM is a localized tumor arising from the epithelial and mesenchymal elements of the mesothelial cells, and does not metastasize. The tumor can attach to serosal surfaces of the intestine and omentum or in the retroperitoneal space, spleen and liver if located in the peritoneal cavity (12). Grossly, the cysts filled with mucinous or gelatinous fluid range from several millimeters to more than 20 cm in diameter (8).

Characteristically, the tumor is composed of a multiple mesothelial-lined cystic structure, with fragile fibrovascular stroma holding the formation together (1), as in our case.

The pathogenesis is controversial. Some authors believe that the lesion is neoplastic, while some favor a reactive process (3, 7). The presence of a history of prior surgery, endometriosis, uterine leiomyoma, and inflammation suggests that it is a peculiar peritoneal reaction to chronic irritation stimuli, with mesothelial cell entrapment, reactive proliferation and cystic formation. In addition, an association of BMPM with familial Mediterranean fever characterized by periodic fever and peritonitis reinforces this assumption (13). Weiss and Tavassoli (1) could not confirm this assumption. The neoplastic origin is based on a slow but progressive growth of untreated lesions, tendency to recurrence, a low incidence of previous abdominal infection, and a high disease-related mortality (1, 14). The great majority of patients being women of reproductive age suggests a key role for female sex hormones in its pathogenesis (7). Our patient had no history of surgery.

Cystic lymphangioma (cystic hygroma), cystic adenomatoid tumor, cystic forms of endosalpingiosis, endometriosis, Müllerian cysts involving the retroperitoneum, and cystic mesonephric duct remnants are included in the differential diagnosis among benign lesions. Malignant lesions include malignant mesothelioma and serous tumors of the peritoneum. In addition to hematoxylin-eosin sections, immunohistochemical staining may be useful in the differential diagnosis (7, 15). Among these, BMPM is most likely to be confused with multilocular cystic lymphangioma; however, cystic lymphangioma is usually seen in males younger than 5 years. Microscopically, lymphoid aggregates and smooth muscle are present in their walls. Immunohistochemistry is also helpful in problematic cases (5, 7). The cystic component in cystic adenomatoid tumor is usually accompanied by a recognizable solid component. There are cases of tumors with mixed features of both. Thus, these two lesions are probably pathogenetically related (3). Positive staining with calretinin, vimentin and Wilms' tumor antigen, and negative staining for Ber-EP-4, carcinoembryonic antigen (CEA) and CD15 indicate mesothelial lineage (8). Malignant tumors mimicking BMPM show significant cytologic atypia or increased mitotic activity, at least focally within the lesion (7). In addition, malignant mesothelioma grossly appears as multiple plaques or nodules, which are almost always associated with ascites (8).

Tumor recurrence is reported in about half of the patients, even when all gross and visible tumor tissues have been removed (1, 8). Thus, routine follow-up imaging is required in patients after operation (16). The prognosis is excellent and the only death that has been reported in the literature occurred in a patient who refused to undergo resection 12 years after diagnosis (1). A 36-year-old woman was followed for 10 years, and developed an aggressive, diffuse malignant mesothelioma after six surgical procedures (17).

Surgery is the only effective treatment. Aggressive surgical approaches including cytoreductive surgery with peritonectomy are recommended (18). However, sclerosive therapy with tetracycli-

ne, continuous hyperthermic peritoneal perfusion with cisplatin and peritonectomy with intraperitoneal chemotherapy have been attempted in individual cases with varied degrees of success (7).

Pre- and peri-operative recognition of BMPM is difficult but important for proper surgical management. Although approximately 130 cases have been described in the literature, publication of case reports on BMPM is required to better define this entity.

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