

# Giant Benign Multicystic Peritoneal Mesothelioma: Case Report

## Dev Benign Multikistik Peritoneal Mezotelyoma

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**ABSTRACT** Benign multicystic peritoneal mesothelioma (BMPM), which must be taken into account in intraabdominal cysts, is an important pathology due to the potential of local recurrences and malignant transformation. Although BMPM occurs mostly in women in their reproductive age, small cases have been described in male patients. We hereby present the radiological and histopathological findings of a 45-year-old male patient who was admitted with the complaints of discomfort, swelling and abdominal pain. A giant multilocular cyst of 35x20 cm in size, revealed pre-operatively via computed tomography, was removed surgically. Generally single-rowed and occasionally 2-3 rowed cuboidal epithelium were observed in the histopathological assessment of the cyst which contained multiple semi-transparent vesicular structures at a diameter of 0.3-1 cm. Cytokeratin, Calretinin and epithelial membran antigen (EMA) staining confirmed its mesothelial origin immunohistochemically and the diagnosis of BMPM was made. Giant cyst removed via a successful operation and no recurrences were detected at the 32-months post-operative period. In BMPM cases differential diagnosis versus other cystic diseases can only be based on histologically. This is critical importance to clinical management.

**Key Words:** Mesothelioma, cystic; peritoneum

**ÖZET** İntraabdominal kistlerde akılda bulundurulması gereken benign multikistik peritoneal mezotelyoma (BMPM) lokal nüks ve malign transformasyon olasılığı nedeni ile önemli bir patolojidir. Daha çok kadınlarda ve reproduktif dönemde görülmesine karşın az sayıda erkek hasta bildirilmiştir. Biz 45 yaşında huzursuzluk, şişlik ve karın ağrısı şikayeti ile başvuran erkek hastayı sunuyoruz. Preoperatif bilgisayarlı tomografide multiloküle görünümdeki 35x20 cm çaplı dev kistik yapı cerrahi olarak çıkarıldı. Üzerinde 0,3-1 cm çaplı çok sayıda yarısaydam veziküler yapılar içeren kistin histopatolojik incelemesinde genellikle tek sıralı, yer yer 2-3 sıralı küboidal epitel görüldü. İmmünohistokimyasal olarak Keratin, Kalretinin ve Epitelyal Membran Antijen (EMA) boyaları ile mezotelyal orijini doğrulandı ve BMPM tanısı kondu. Dev kist başarılı bir operasyonla çıkarıldı ve postoperatif 32. ayında nüks tespit edilmedi. Benign multikistik peritoneal mezotelyoma olgularında diğer kistlere karşı ayırıcı tanı yalnızca histoloji temelinde yapılabilir. Bu klinik tedavi yönetiminde hayattır.

**Anahtar Kelimeler:** Mezotelyoma, kistik; periton

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**B**enign multicystic peritoneal mesothelioma (BMPM) which mostly occurs in women in their reproductive age is a rare type of tumor arising from the peritoneal mesothelium that covers the abdomen and the pelvic cavity.<sup>1</sup> It was first described by Plaut in 1928 and it was first named as BMPM by Mennemeyer and Smith in 1979.<sup>2,3</sup>

Although the etiology of BMPM is unclear, it has been reported that previous abdominal surgery, endometriosis and pelvic inflammatory disease may play a role in the etiology of the disease.<sup>4,5</sup> BMPM has been reported to date about 140 cases in the literature, small cases have been described in male patients.<sup>6</sup>

Surgery is mandatory for both diagnosis and treatment. We hereby present the clinical, radiological and pathological findings of a case with giant BMPM which has a certain extent in the literature. We believe this to be the first case of BMPM with the largest diameter of the peritoneum reported in the Turkish literature.

## CASE REPORT

A 45-year-old male patient presented to the surgery clinic with the complaints of abdominal pain, swelling and uneasiness that lasted for a few months. In his anamnesis, he had no previous operation and no irritable bowel syndrome. There was nothing remarkable in his family history. On physical examination, there was abdominal distention and tenderness. He had no loss of appetite or weight loss. All the laboratory results, including complete blood count, liver and renal function tests, all biochemical values, and markers such as CA-125 and CA-19-9 were within normal ranges. On preoperative computed tomography (CT) examination, in the middle and lower portion of the abdomen, a giant multilocular cystic mass with no fatty planes between surrounding tissues including septas with mild contrast enhancement displacing the intestines to posterior and superior was revealed (Figure 1).

The patient underwent an exploration with the prediagnosis of mesenteric cyst. On exploration, a large cystic mass with grape-like vesicular structures partly adherent to the omentum was completely resected and the specimen was analyzed histopathologically.

On gross inspection, the specimen had a multicystic appearance with dimensions of 35x20 cm having semi-transparent vesicle cysts on superior in 0.3-1 cm diameters, and pale yellow serous fluid

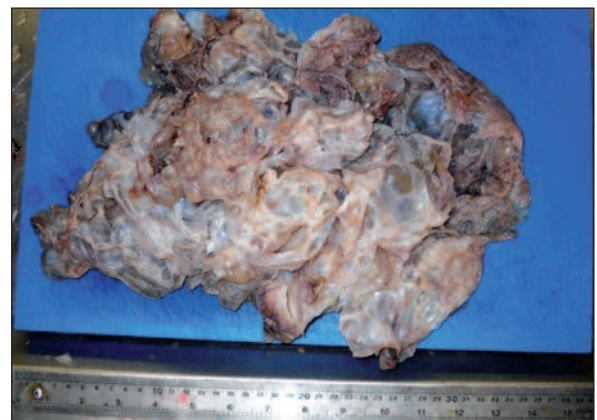


**FIGURE 1:** Abdominal CT showed a large, septated multilocular cystic mass with little enhancement by contrast material.

discharge at incision site. The cyst wall had a multilayered structure with a diameter of 0.5 cm at its widest part (Figure 2).

On microscopic evaluation, adjoined cystic cavities were observed that possessed a thin connective stroma, mostly paved with single row cuboidal epithelium, and in a few areas with 2-3 rows of cuboidal epithelium. There were no cytological atypia, mitosis and stromal invasion in the epithelia of the cyst. There were also lymphoid aggregates in the poor adipose tissue (Figure 3,4).

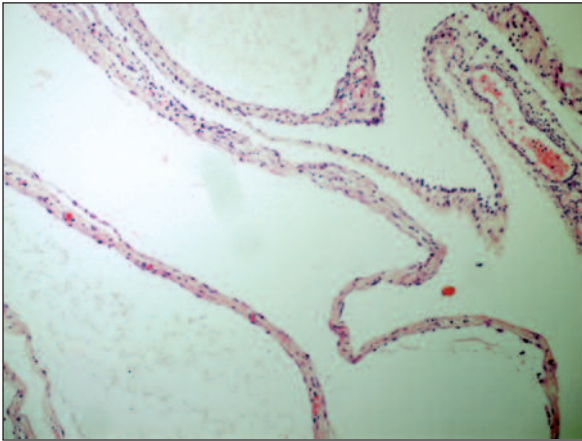
The mesothelial origin was confirmed by the immunohistochemical staining of the cyst epithelium with cytokeratin, calretinin and EMA (epithelial membran antigen) (Figure 5A-C). However, a positive staining was observed with



**FIGURE 2:** Gross photograph showed multilocular cystic tumor, 35 cm in diameter. Opening the surgical specimen revealed that the tumor had some septums and contained yellowish transparent fluid.

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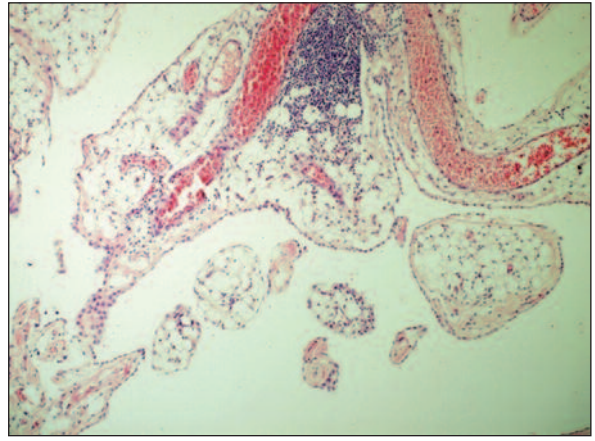
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**FIGURE 3:** Light microscopy showed cystic spaces lined by flat cells and intervening connective tissue stroma (H&E, X20).

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**FIGURE 4:** Scattered lymphoid aggregates observed in the lipomatous stroma. (H&E, X20).

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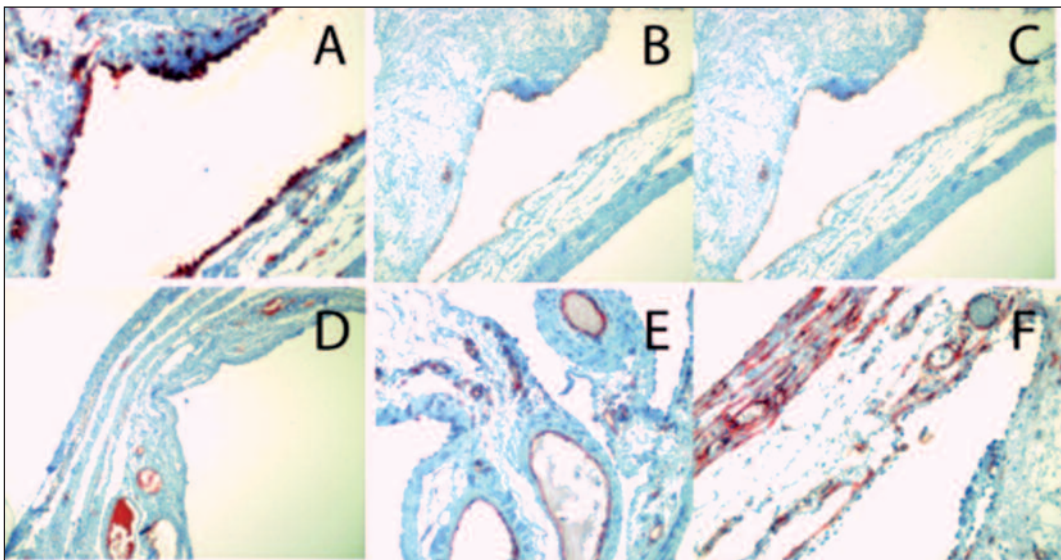
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factor VIII and CD 31 only in the vessel walls (Figure 5D,E). The SMA (smooth muscle actin) positivity was obtained in the cyst wall (Figure 5F). According to the microscopic findings and immunohistochemical staining, the case was reported with the diagnosis of BMPM, and discharged from the clinic with full recovery after 3 days. The patient is currently alive without metastasis at 32 months postoperatively.

Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

## DISCUSSION

Benign multicystic mesothelioma particularly originates from the pelvic retroperitoneum and tends to locate on the serosal surface of pelvic viscera.<sup>1</sup> Histopathologically, the tumor is composed of nu-



**FIGURE 5:** Immunohistochemical findings are showing mesothelial origin. (A-B-C); strong membranous immunoreactivity for cytokeratin, calretinin and EMA in cells lining the cysts. (D-E); Factor VIII and CD 31 stains are excluding cystic lymphangioma. Positivity for factor. (F); immunoreactivity for SMA in the wall of the cyst. (X20).

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merous adjacent cystic structures paved with mesothelial cells having a thin fibrovascular stroma. On macroscopic evaluation, there are cysts filled with mucinous-gelatinous fluid with diameters ranging from few millimeters to 20 cm or above.<sup>1,7</sup> In our case the resection specimen had a multicystic appearance with dimensions of 35x20 cm.

This benign tumor with pathogenesis of inflammation, endometriosis and previous surgeries may lead to mesothelial proliferation and cyst formation as a result of peritoneal reaction causing chronic irritation.<sup>1,4-6</sup> Although the patient had no previous operation history he was admitted with the complaints of discomfort, swelling and abdominal pain.

The reason for encountering a higher frequency of BMPM in the reproductive age may be related to the estrogen and progesterone receptors, and supports the key role of female sex hormones.<sup>8</sup> However, there are studies reporting BMPM in children and males.<sup>1,6,7,9,10</sup> Although BMPM generally presents with pelvic mass findings, it should be kept in mind that BMPM also presents with the symptoms of an abdominal mass, swelling, weight loss and scrotal herniation in men.<sup>7,10</sup>

Although BMPM does not cause metastasis, confirming its diagnosis prior to operation provides additional information to the surgeon for the requirement of complete removal of the cyst because of the risk for recurrence and possible malignant transformation.<sup>11,12</sup> However, the diagnosis of BMPM is difficult despite the clinical assessment, measurement of tumor markers, and utilization of imaging techniques. In spite of the frequent current use of advanced radiological imaging techniques, the differential diagnosis of BMPM is difficult due to the possibility of cystic lesions such as mesenteric cysts, lymphangiomas, pancreatic pseudocysts, endometriomas, peritoneal inclusion cysts, cystic adenomatoid tumors, hydatid cysts, cystic teratomas and urogenital cysts.<sup>6</sup> However, detection of abundant numbers of mesothelial cells on cytological evaluation of peritoneal irrigation fluid may assist to perform preoperative diagnosis.<sup>13</sup>

A precise diagnosis based on imaging findings alone is not possible. Furthermore, distinguishing a benign from a malignant process as well as a primary from a metastatic process is also challenging. Therefore, the definitive diagnosis of peritoneal mesothelioma depends on histologic and immunohistochemical examination. Immunohistochemistry uses some cytological markers: calretinin and cytokeratin are used to determine whether the tissue is mesothelial.<sup>6</sup>

The primary pathologic differentiation in multilocular cysts of the peritoneum is between BMPM and cystic lymphangioma. Lymphangioma is mostly encountered in children and males.<sup>1,6,7,9,10</sup> The cystic spaces are lined by a single layer of flattened endothelial cells which are immunoreactive to vascular markers (CD31, CD34, factor VIII). Smooth muscle cells are found in its cyst wall. On the other hand, BMPM is most commonly encountered in women and the cyst wall is paved with mesothelial cells without smooth muscle cells. In our case, sporadic SMA positivity and small lymphoid aggregates in adipose tissue were observed. However, despite the positive immunoreactivity of the cyst epithelium with mesothelial markers (cytokeratin, calretinin), no positive staining was obtained with CD31 and factor VIII.

The differentiation between benign and malignant mesothelioma can be easily performed by the absence of cytological atypia, mitotic activity, stromal infiltration, and gross appearances of plaques and nodules. Even there is no exposure to asbestos, recurrence and malignant transformation can be suggested to be variants of benign and malignant forms of BMPM.<sup>11</sup> Miles et al. reported a recurrence rate as 75% between a period of 1 month to 16 years. Death has been reported in one infant with a cystic diffuse epithelial mesothelioma, and in one patient who refused resection 12 years after the diagnosis.<sup>1</sup>

Despite the lack of a consensus in the treatment and follow-up of BMPM, one group of surgeons minimizes the recurrence rate by performing complete peritonectomy and cytoreduction.<sup>14</sup> Another group of surgeons, however, support an aggressive treatment method com-

prising chemotherapy and radiotherapy in addition to the resection due to the presence of the possibility of recurrence and malignant transformation.<sup>15</sup> Moreover, peritonectomy together with intraabdominal tetracycline cisplatin and peritoneal perfusion are among the treatment methods.<sup>8</sup>

Hereby, we presented a case with a large cystic tumor of 35 cm in diameter which was resected by a successful surgical operation, and with no recurrence in postoperative 32 months. This lesion

has a non-specific appearance on imaging which does not permit differential diagnosis from other cystic lesions and always requires histological evaluation. Histological examination, accurately with the aid of immunohistochemical techniques, is therefore essential for diagnosis.

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