Case Report

Dig Surg 1993;10:203–207

Cystic Mesothelioma of the Peritoneum

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Key Words
Mesothelioma, cystic
Peritoneum

Abstract
Cystic mesothelioma is a rare benign tumor of the peritoneum characterized by multiple free and attached cysts of the abdominal or pelvic cavity. The present study concerns a single 36-year-old female patient with cystic mesothelioma. Clinical, radiological and pathologic features are discussed.

Introduction

Cystic mesothelioma (CM) is a rare benign tumor of the peritoneum characterized by multiple free and attached cysts of the abdominal or pelvic cavity. The tumor was originally described by Plant [1] in 1928 as loose cysts of the pelvis, incidentally discovered during surgery for uterine leiomyomas.

In 1979, Mennemeyer and Smith [2] presented the first ultrastructurally documented CM in a patient. Since then, approximately 100 cases of CM have been described in the literature. Excluding one case report [3] the lesion has been described exclusively in the abdomen. The exception is a 37-year-old woman, described by Ball et al. [3], in whom the CM originated from the pleural cavity. It had all the pathologic features (including immunohistochemistry and electron microscopy) of the peritoneal mesothelial lesions.

In the present report, the case of a 36-year-old female patient with CM is described, and clinical, radiological and pathologic features are discussed.

Case Report

A healthy, 36-year-old woman, complaining of vague abdominal pains and an increase in abdominal girth, was referred for an abdominal examination to the ultrasound department of Carmel Hospital, Haifa. The medical history was unremarkable, excluding three cesarian sections. No intraperitoneal surgery had been performed.

The ultrasound examination revealed multiple cystic structures filling the entire abdominal cavity as well as the pelvis. The cysts were of variable sizes (fig. 1). A computerized tomography (CT) scan demonstrated the findings as well (fig. 2); however, the septations, so clearly seen by ultrasound, were less prominent with this imaging procedure.

Abdominal surgery was performed, and the omentum was found to contain multiple cystic structures (fig. 3). The round, grape-like locules ranged from 0.5 to 5 cm in diameter and revealed a thin, translucent, pink-to-white wall, containing serous to serosanguineous fluid. All cystic structures, which measured 23 × 20 × 7 cm, were excised. No mucocele was found in the appendix, and no mucinous cystadenocarcinoma could be detected either in the appendix or in the ovaries.

Follow-Up

Two months after surgery a first follow-up ultrasound examination was negative. A second ultrasound, performed 6 months after surgery, was also normal. However, in the third ultrasound examina-

Received:
May 6, 1993
Accepted:
October 4, 1993

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0253–4886/93/0104–0203
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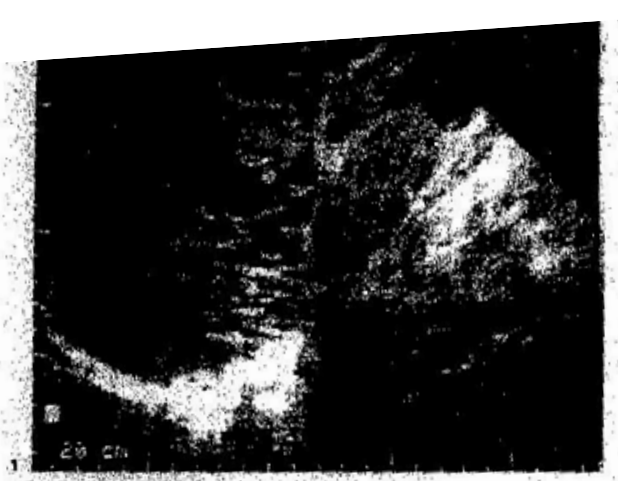

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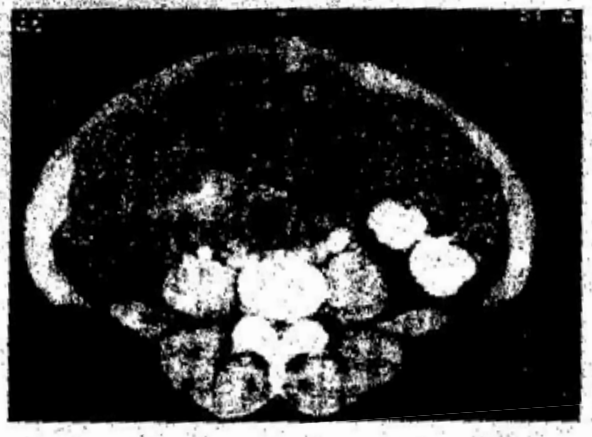

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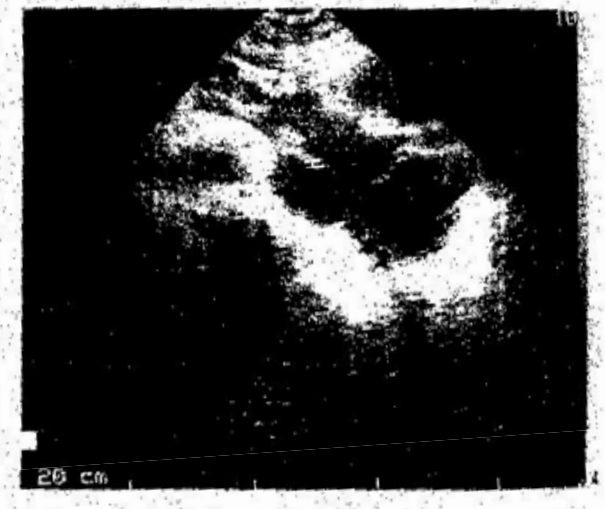

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Fig. 1. Longitudinal ultrasound scan through the right upper abdomen. Several cysts are identified between the liver (L) and the kidney (K).

Fig. 2. CT scan through lower abdomen. The whole abdominal cavity is filled with the lesion (CM). The septations between the cysts are less prominent compared to the ultrasound scan (fig. 1). B = Bowel loop.

Fig. 3. The excised multicystic omental mass (23 × 20 × 7 cm).

Fig. 4. A multiseptated cystic mass in the pelvis detected by ultrasound about a year after surgery.

tion, performed about a year after surgery, a number of cysts could be identified in the pelvis (fig. 4), as well as between the right kidney and the liver (Morisson's pouch), and between the left kidney and the spleen. These findings suggest recurrence of the CM.

Microscopic Findings

The lesions were composed of multiple cysts of different sizes separated by variable amounts of stroma (fig. 5). Flat to cuboidal mesothelial cells lined the locules, and foci of squamous metaplasia were seen. The fibroblastic stroma showed areas of chronic inflammatory infiltration, old and fresh hemorrhages and fibrin deposition (fig. 6).

Immunostaining for cytokeratin revealed strong positivity within the cells lining the cysts, and electron-microscopic examination of the cyst-lining cells revealed long, thin microvilli on their luminal surface and numerous lateral desmosomes which formed connections among the cells. A basal lamina separated the mesothelial cells from the subjacent stroma.

Fig. 5. Multicystic mesothelioma composed of numerous mesothelial lined cysts embedded in a fibrovascular stroma. Hematoxylin-eosin. × 25.

Fig. 6. Fibroblastic stroma showing areas of chronic inflammatory infiltration, old and fresh hemorrhages and fibrin deposition. Hematoxylin-eosin. × 125.

Discussion

CM is a rare tumor, occurring most commonly in women of reproductive age, with a 5:1 female-to-male ratio. The average age in females is 38 years, with a range between 20 and 73 years [4], while for men, the average age is 41.5 years [5]. CM has been described in children as well [4, 6] and even in a newborn [7]. Lately [8] a case of a benign CM has been described in a 79-year-old women.

The tumor can be intra- or retroperitoneal, the latter being very uncommon and considered potentially malignant. A cystic retroperitoneal perinephric mesothelioma was described by Furuta et al. [9]. A 70-year-old man, complaining of discomfort in the right flank, was examined. CT revealed, in the right retroperitoneal space, multiple cystic masses which infiltrated the kidney and the psoas muscles. The patient was operated; the total mass was 16 × 10 × 8 cm in size and 630 g in weight. The histological diagnosis was diffuse benign multicystic mesothelioma. Another patient with CM of the retroperitoneum is also presented by Li et al. [10].

In describing 5 patients with CM, O'Neil et al. [11] concluded that CM of the peritoneum has a nonspecific multilocular cystic appearance on images, which does not permit it to be differentiated from other cystic lesions.

The main differential diagnosis of CM is lymphangioma. In fact, it is assumed that many cases, reported in the past as CM, were actually lymphangiomas. Suh and Choi [12] presented a 53-year-old woman who had abdominal pain and palpable mass. On operation the mass invaded the peritoneum in the region of the appendix. Electron-microscopic examination confirmed that the cells were mesothelial in origin. The authors discuss that lesions of CM can mimic cystic lymphangioma of the abdomen.

Pseudomyxoma peritonei is another lesion that might mimic CM. This is a rare lesion originating from a ruptured mucinous tumor of the appendix or pseudomucinous ovarian cyst. These two tumors coexist not uncommonly [13]. The rupture causes seeding of the omentum and the peritoneum with *mucinous* implants. Sonography and CT examinations can demonstrate multilocular cysts which are difficult to be distinguished from CM.

In a case report of CM published by Hasan and Sinclair [8], the CT examination showed calcification within the mass. In that particular case, the location of the mass and the existence of calcification raised a differential diagnosis between adrenal carcinoma and teratoma.

Other possibilities in the differential diagnosis list should include cystic mesonephric remnants and epithelial inclusion cysts [14].

Occasionally extraovarian peritoneal implants of serous borderline tumors of the ovary may mimic CM. 20–46% of serous borderline tumors are associated with such implants.

In most cases the peritoneum is studded with multiple nodules. However, on occasion, peritoneal cysts can be seen [15] especially on the broad ligament surface.

These extraovarian borderline serous tumors should be differentiated from primary peritoneal serous epithelial lesions that arise from the peritoneal mesothelium. The primary peritoneal tumors are usually of small size (few millimeters in diameter [16]) and are unlikely to be con-

... microscopically with CM.

Surgery is considered to be the treatment of choice for CM. However, Hanukoglu et al. [6] emphasize the importance of a conservative approach to this lesion in childhood. They described an 11-year-old boy in whom a benign CM occupied the entire peritoneal cavity.

Benson and Williams [17] presented a patient in whom several unsuccessful attempts of surgical resection had been performed. The patient subsequently underwent a combination of conservative surgical resection and sclerosive therapy with tetracycline. No recurrence was noticed during a 4-year period of follow-up.

Hidvegi et al. [18] described a 25-year-old woman with an 8-year history of 5 operations because of recurrent tumor.

Iversen et al. [19] reported a 27-year-old woman who had been operated on for multicystic benign mesothelioma, with free-floating, thin-walled cysts in the abdominal cavity. After removal of all visible cysts by laparotomy, the patient was well for 29 years, at which time she required an operation for cholecystitis and gallbladder stones. During the operation, the whole peritoneum was found to be covered with small cysts, lined by mesothelial cells. The authors discuss whether such a lesion is a benign tumor or should rather be otherwise classified.

No consistent etiologic factor for CM has been found [20, 21], but the high rate of these lesions in women has inspired speculation that one of the risk factors of CM is endometriosis [22]. The assumption is that peritoneal irritation plays an important role in the development of the lesion. Others [4] consider CM as a hamartomatous lesion due to an abnormal development of the peritoneal layer. Villaschi et al. [23] described 3 women with CM who had previous abdominal surgery. The authors suggest that an earlier laparotomy may be a precursor to CM.

This theory has been rejected by Weiss and Tavassoli [14]. They published a series of 37 cases with multicystic mesothelioma. Very few patients in their study had previous surgery, and no one had proven abdominal infection. Out of 25 patients, who were followed up, 2 had died because of the tumor. Due to that fact the authors consider CM as a neoplastic lesion.

Ball et al. [3] consider the lesion as a reactive process. The assumption is based on the fact that in their case of CM there was continuity of the lesion with normal surrounding mesothelium. Another suggested factor for CM is abdominal trauma [24].

Unlike malignant mesothelioma, there is no known association between the benign CM and asbestos exposure. Nevertheless, the patient reported by Ball et al. [3] (having CM of the pleural cavity) had been exposed to asbestos 20 years prior to the diagnosis. Kjellevold et al. [25] had a case of peritoneal CM with a past history of asbestos exposure.

Clinical Presentation

The symptoms of the disease are usually abdominal discomfort and pain, increase in abdominal girth or symptoms related to local pressure on certain organs [2, 6, 9]. Other manifestations include early satiety, urinary urgency and frequency, and dyspareunia [11]. On physical examination, a tender mass or a vague pelvic fullness may be palpated [11, 21].

CM may mimic other acute abdominal conditions, as in a 40-year-old woman with signs of acute appendicitis, described by Betta et al. [26].

Radiological Findings

A large multicystic mass may be seen on ultrasound or CT of the abdomen, done for evaluation of a tender lower abdominal mass [5, 7, 27]. On ultrasound, the cysts are uniformly anechoic and without calcification. A single case report describing a calcification in a case of CM has been already mentioned before [8].

Magnetic resonance imaging may add clarity in determining the position of the mass relative to the bowel, but it is no more sensitive for lesion detection than CT. Magnetic resonance imaging signal characteristics of the fluid in the mass can help in excluding the presence of fat or hemorrhage [11].

Pathologic Findings

Three well-characterized types of mesotheliomas have been reported in the peritoneum: adenomatoid tumors (benign mesothelioma), CM and malignant tubulopapillary mesotheliomas [24, 28]. The common adenomatoid tumors are small asymptomatic lesions, found incidentally at laparotomy. They appear primarily in the epididymis and in the uterus or fallopian tubes. Palacios et al. [29] presented a 38-year-old woman with cystic adenomatoid tumors of the uterus that histologically, immunohistochemically and ultrastructurally closely resemble those of benign CM.

Malignant mesotheliomas are highly lethal tumors that occur predominantly in men [30]. They have been causally related to asbestos exposure.

Histopathologically, the cysts of benign mesothelioma are lined with a single layer of benign mesothelial cells, separated by loose connective tissue. They have a 'hob-

appearance [31]. Electron microscopy shows cystic spaces, lined by cuboidal or flattened cells with numerous microvilli and well-formed desmosomal junctions.

Treatment

Treatment of benign cystic mesothelioma is by surgical resection. Complete resection should be performed, and recurrences should be treated with repeated surgery. However, in childhood a conservative approach has been suggested [5].

The prognosis of CM is good. However, Weiss and Tavassoli [14] found either adenomatoid change or squamous metaplasia in about one third of their 37 patients with CM. In their series they had two deaths related to the lesion. In one case transition to conventional mesothelioma was found. Morbidity is related to local spread and recurrence and is attributed to local spread of the lesion.

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