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Cystic Peritoneal Mesothelioma: Report of a Case

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Abstract: Peritoneal mesothelioma is a rare disease, especially when it arises in a cystic form with tardive and often nonspecific symptoms. While diffuse neoplasms have an unfavorable prognosis, cystic forms are usually benign. An accurate diagnosis can only be made only with electron microscopy and immunohistochemical studies. A 92-year-old woman with an ultrastructurally ascertained cystic peritoneal mesothelioma was admitted to the hospital's emergency ward, and was considered to be unusual because of the size of the mass and the patient's age. A review of the literature is made, and the clinical and diagnostic aspects of this disease are also discussed.

Key Words: peritoneal neoplasm, cystic peritoneal mesothelioma

Introduction

Malignant mesothelioma is a rare disease with an incidence of 0.9–1.0 per million persons per year.[1] Diffused malignant mesothelioma (DMM) is more frequently observed than cystic mesothelioma (CM). There have been few reports in the literature of cases that occur at a very advanced age and that present with acute abdomen.

Case Report

A 92-year old woman was admitted to our hospital presenting with abdominal swelling. She reported a family history of neoplastic disease, since her brother and father both died of pharyngeal carcinoma. During the preceding 5 months, she had suffered abdominal pain. Ultrasonography of the abdomen revealed a partially solid mass in the left abdomen, most likely beginning in the adnexa. Magnetic resonance imaging confirmed the presence of a 147 × 146 × 121 mm formation, believed to originate in the ovaries but with an absence of ascites (Fig. 1). In the subsequent months, the patient also complained of asthenia.

Following a spell of sudden fainting and the evacuation of dark and pitchy stools, she was admitted to the hospital in a poor general condition. A physical examination revealed a very distended abdomen, and the abdominal quadrants were sore. She had numerous dilated superficial veins and marked enterocolic tympanism elicited by percussion. Routine laboratory tests showed severe anemia: RBC 2340000/mm^3, hemoglobin 5.6 g/dl with no alterations of either the number or the formula of the WBCs.

The patient immediately underwent surgery which consisted of a midline laparotomy. A very large mass measuring a maximum of 30 cm in diameter was found which displaced the small intestinal loops and the transverse colon to the right, while the stomach was dilated and shifted anteriorly downwards. The formation was highly vascularized and extended below the liver, the mesentery, and above the pancreas. An en bloc resection of the mass was thus performed.

A histological examination revealed a compact, whitish-grey, translucid, soft, and elastic tissue composed of fusiform cells with oval and dark nuclei and scarce cytoplasm, clustered together. Mitodes were 1–2/10 H.P.F. There were highly cellular areas close to areas with scanty cells with mixoid modification, collagen arranged in tight bands or plaque-like masses, and cystic spaces surrounded by a flattened cubic epithelium which was keratin-positive (Fig. 2). In addition, immunohistochemical staining was positive for vimentin, keratin, and a histiocytic marker (Fig. 3). The diagnosis was cystic peritoneal mesothelioma.

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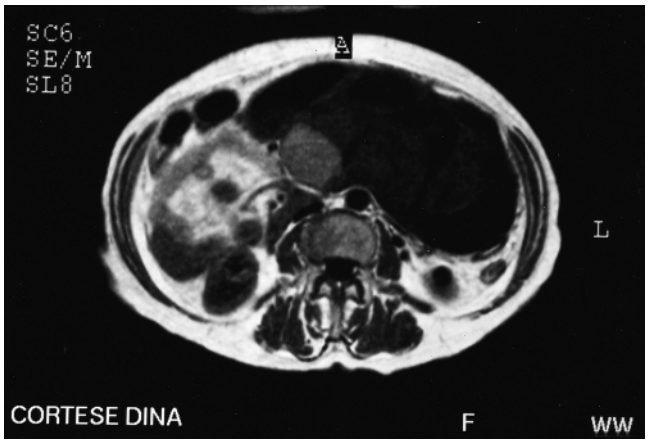

Fig. 1. A partially solid mass measuring 147 × 146 × 121 mm in diameter in the left abdomen

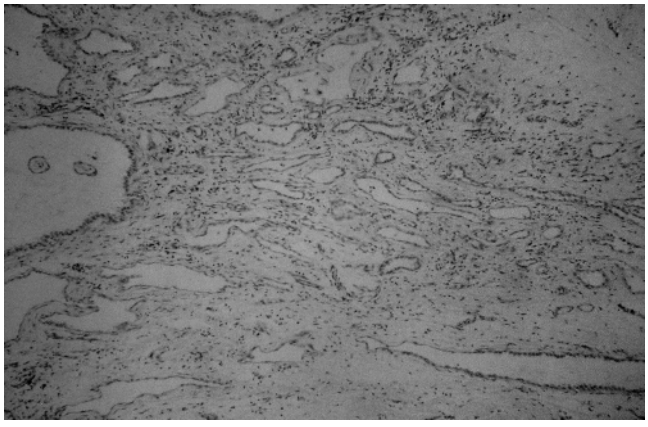

Fig. 2. Multiple cysts lined by a layer of flattened or cuboid mesothelial cells with a little chronic inflammatory infiltration

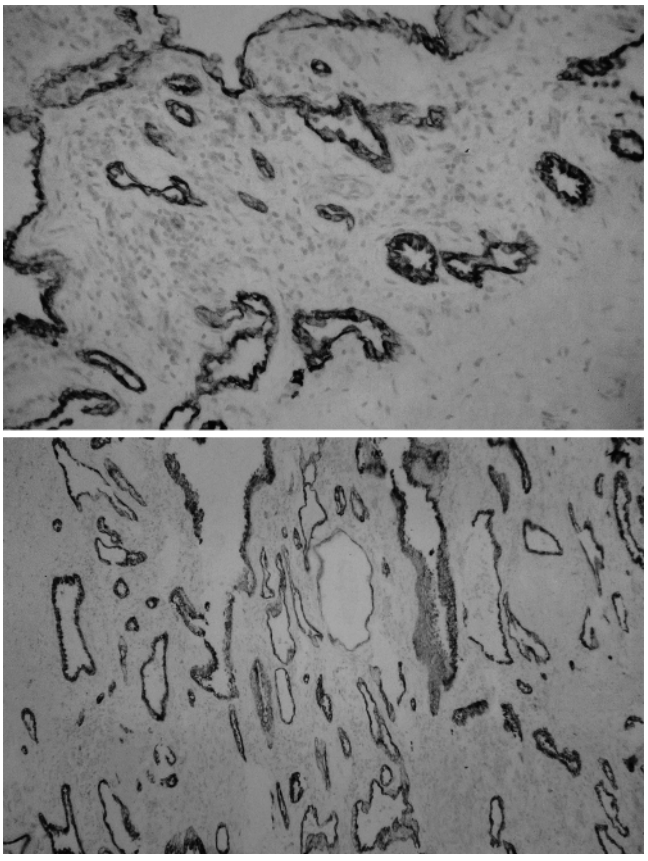

Fig. 3a,b. Immunohistochemical study. **a** The cells lining cysts are keratin-positive. **b** The same cells are also HBME 1-positive while they are negative for specific markers for endothelial cells (factor VIII-related antigen, CD31, and CD34)

The patient's postoperative course was uneventful and she was discharged on the 13th postoperative day. After 12 months, she is in fairly good condition with no sign of recurrent disease.

Discussion

Mesotheliomas originate from cells that cover the serous cavity. Those that arise in the peritoneum account for only 20%.[2] From a clinical and morphological point of view, peritoneal mesotheliomas may be either diffuse or localized. The former types originating in the peritoneum include the following: (1) diffuse malignant mesothelioma (DMM) with an unfavorable prognosis; (2) well-differentiated papillary mesothelioma (wDPM); and (3) cystic mesothelioma (CM) which is a benign tumor, even though recurrence may be observed in 50% of the cases treated surgically.[2,3]

Macroscopically, the DMM and wDPM types may be observed in the form of solid whitish plaques or nodules that can be limited or can spread to the entire peritoneum. CM, instead, presents as translucid cysts, separated by well-defined fibrous septa. In time, these lesions tend to come together and may invade the surrounding organs.

From an ultrastructural point of view, CM may consist of cuboidal cells in which mitoses are either absent or rare. On the contrary, DMM shows an atypical cytology with mitoses, necrosis, and vascular proliferation associated with infiltration of both lymphocytes and the surrounding tissues. In this latter form, the cells may be epithelial (75%), sarcomatoid (2.4%), and of a biphasic or mixed type (22%), forming tubulopapillary structures that are difficult to distinguish from an adenocarcinoma.[2]

The most common mesothelial tumor is DMM, whereas CM and wDPM are more rare. Since 1979, when Mennemeyer and Smith described the first case of CM,[4] approximately 80 cases have been reported in the literature. The age of the patients affected by this tumor

ranges between 2 and 92 years. While DMM is more frequent in males (two thirds of cases) with a greater frequency between the ages of 45 and 64 years, CM is more commonly observed in young women (male/female ratio: 1:5).[5] According to Katsube et al., the average age at diagnosis for CM is 37.6 years.[6]

Regarding the etiopathogenesis of these tumors, there are clear differences between the malignant form and CM. While various authors agree that the occurrence of mesotheliomas is related to prolonged exposure to asbestos, no precise etiological factors have yet been identified regarding the cystic form.[6–8]

Peritoneal mesotheliomas are generally asymptomatic until the tumor extends into the surrounding tissues and reaches a considerable size with the onset of nonspecific symptoms. Ascites, observed in 90% of the cases, is the most common clinical finding in DMM, while the presence of a palpable abdominal and/or pelvic mass is more typical in CM.[4,9]

Although the tumor may remain silent in the abdominal cavity, it may invade such underlying organs such as the omentum, peritoneal serosa, abdomen, pelvis and, more rarely, even the retroperitoneum.[10] As in the case described here, the first sign of the lesion was revealed only a few months before the patient underwent surgery, which was performed in an emergency situation because of the large retroperitoneal mass that occupied and displaced both the small intestinal loops and the transverse colon which closely adhered to all the retroperitoneal structures, while also demonstrating both compression and invasion of the posterior stomach wall.

The diagnosis of peritoneal mesothelioma can be made only after surgery. In fact, even with the currently available techniques (ultrasonography, computed tomography scans, and magnetic resonance imaging), a differential diagnosis cannot be made with other neoplastic and inflammatory diseases that arise in this anatomical area.[10,11]

As already stated, the surgical techniques available for a definitive diagnosis of peritoneal mesothelioma include laparoscopy and laparotomy, especially in emergency cases, in order to explore the abdominal cavity and obtain biopsy samples. The diagnosis is made by electron microscopy and immunohistochemistry using antibodies for cytokeratin, EMA, and vimentin. In fact, optic microscopy alone cannot distinguish either DMMs from metastatic adenocarcinomas, or CMs from cystic lymphangiomas.[4,12]

Surgery is the only effective treatment for benign CM, consisting of a complete removal of the lesion when possible. Because these tumors have a prevalently benign course, adjuvant radio- and chemotherapy are not indicated.[4,6,9,10] The prognosis is excellent and, in spite of a significant rate of local recurrence (in 50% of the cases), no cases of distant metastasis have been reported in the literature[4] with the exception of one patient who died of complications related to local recurrence, 12 years after refusing surgery.[13]

The case described herein is considered to be unusual because of the rare retroperitoneal localization, the patient's age which is not typical for CM, and the combined emergency surgery situation. An analysis of the literature shows that, because CM is a benign disease, highly aggressive procedures that involve the removal of vital organs should not be conducted but instead be limited to a partial removal, while postponing the treatment for possible recurrence until a later time.

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