Benign cystic mesothelioma presenting as a huge pelvic mass - a case report

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Summary

Benign cystic mesothelioma is an extremely rare peritoneal tumor. It is reported in women of childbeering age but also in male it can be found; and it need a very carefully differential diagnosis between benign and malign neoplasia for the choose of adeguate therapy.

A 25-year-old was treated in our clinic for a gigantic cystic mass (diametros 25 x 22 x 3) conteining many cysts of different diameters [1-7]. All tumoral markers were withing normal renge. The surgical treatment consisted of radical excision of the mass with good prognosis.

Introduction

Mennemeyer and Smith described this extremely rare tumor in 1979 [1]. Benign cystic mesothelioma (BCM), also called "multicystic peritoneal mesothelioma", "cystic mesothelioma of the peritoneum", "multilocular cyst of the peritoneum", "multicystic mesothelioma," and "multilocular peritoneal inclusion cyst", is an uncommon lesion of the peritoneum characterized by the formation of multiple, thin-walled, multilocular cysts that frequently produce large, intraabdominal masses, and often recur after surgical resection.

Retroperitoneal cystic mesotheliomas are rare tumors that must be differentiated from a variety of cystic abdominal masses. Approximately 130 cases of BCM have been reported in the literature since 1998 [2]. These tumors occur most commonly in women of childbearing age, although several cases have been reported in men [3].

BCM is primarily reactive or neoplastic in nature, but the fact that the great majority of patients are women of reproductive age suggests a key role for female sex hormones in the pathogenesis. Cysts are most often located on the surfaces of the ovaries, large bowel, fallopian tubes, cul-de-sac, and pelvic sidewalls. It has been difficult to estimate the size of these tumors, either because of cyst rupture and collapse during resection or because some reported measurements appear to have been based on the size of individual locules within a multilocular cyst, rather than the entire lesion. The cysts can range in size from 0.3 to 15 cm [4].

Case Report

A 25-year-old woman was referred to our clinic suffering from abdominal distention, lower abdominal pain and a palpable mass at the lower abdominal wall, which had gradually increased in the previous two months. The patient had a history of four spontaneous vaginal deliveries, regular menstruations, and no history of dysmenorrhea, chronic pelvic pain, pelvic inflammatory disease or endometriosis. Additionally, she had not had any operations. The patient was referred to our clinic with the possibility of diagnosis of bilateral ovarian serous cystadenocarcinoma. After a detailed physical examination, we found a palpable mass filling the lower abdomen and reaching the level of the umbilicus. Ultrasonography demonstrated a gigantic cystic mass containing many cysts in varying diameters from 1 to 7 cm surrounding the uterus (Figure 1). The mass showed multiple thin septations. However, the ovaries could not be demonstrated separately. At first glance, the case looked like ovarian malignancy but ascites was not present and the analysis for tumor markers showed a slightly elevated CA125 level at 45 IU/ml. All other tumor markers such as β-hCG, alpha-fetoprotein, CA15-3, CA19-9, carcinoembryonic antigen, and lactate dehydrogenase were within normal range. Abdominal computed tomography (CT) scanning showed a lesion that filled the whole inferior half of the abdomen up to the level of the umbilicus surrounding the uterus and causing upward displacement of the intestines. It was a multilocular giant cystic lesion that had regular borders and contained multiple tiny septations. We opted to perform laparotomy. During the operation, we observed a cystic mass that showed adhesion to the omentum, ovaries, uterus, large bowel, fallopian tubes, cul-de-sac and peritoneum. The mass filled the lower abdomen and consisted of hundreds of serous, translucent cysts in varying diameters from 0.5 to 6 cm (Figure 2). Frozen section analysis revealed benign cystic mesothelioma. After adhesiolysis the uterus and ovaries were observed intact and in regular shape. The mass was excised with the omentum and adherent peritoneum. Total mass dimension measured after excision was 25 x 22 x 3 cm.

Final pathologic analysis confirmed the previous diagnosis.

Discussion

Benign cystic mesothelioma has an unknown etiology, but has been associated with trauma, infection and leiomyomatosis disseminated peritoneal, and there is often a history of previous surgery, pelvic inflammatory disease, or endometriosis [5]. These peritoneal tumors may result in pelvic pain, present clinically as a mass, such as in our case, or can be discovered incidentally during surgery for other reasons. Association between asbestos exposure and BCM has not been definitely established, unlike the conclusive link between asbestos and malignant mesothelioma.

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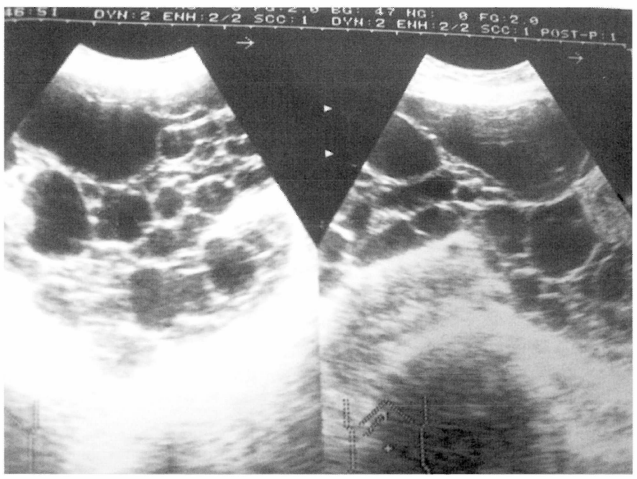
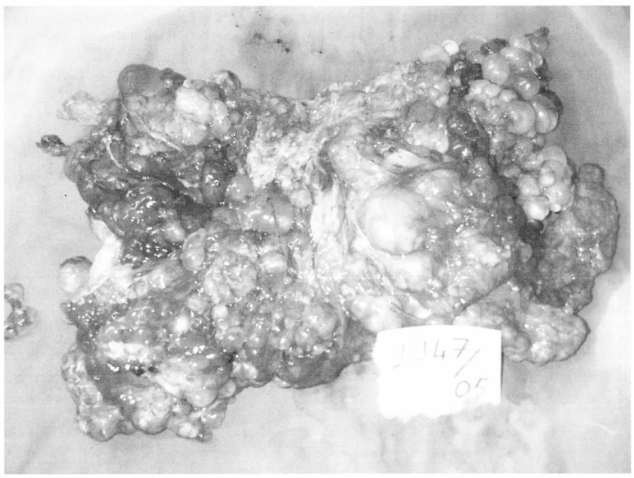
Fig. 1





Fig

Figure 1. — The ultrasonographic appearance of benign cystic mesothelioma.
Figure 2. — Appearance of the benign cystic mesothelioma mass after removal. Note the multiple translucent cysts.

The main differential diagnosis is with cystic lymphangioma, as morphologically BCM resembles lymphangiomas [6]. Distinction can be made by immunohistochemistry. The mesothelial cells react immunohistochemically for calretinin, keratin and vimentin, whereas there is no staining for FVIII-RA, CA19-9, CA125 and CEA antibodies. In distinguishing BCM from malignant mesothelial tumors, differences in desmin and epithelial membrane antigen (EMA) expression patterns might be useful [7].

Although the name is benign cystic mesothelioma, the natural history of this entity is characterized by a great tendency to local recurrence, which is approximately 50% after an interval of three to 27 months [4, 8]. However, no cases of metastases or malignant degeneration have been reported. Some authors consider BCM as having a reactive origin because of a frequent history of abdominal surgery or inflammation [9, 10]. The tendency of these tumors for recurrence can be explained by persistence of the original inciting factor. Recommended treatment for BCM is complete resection and the prognosis is very good, with only one case of death reported in the literature, which was a patient who refused resection [11]. Although there are some authors reporting good responses to anti-estrogen agent tamoxifen, gonadotrophin-release hormone agonist, and intraperitoneal cisplatin treatment in individual cases [12-14], adjuvant radiotherapy or chemotherapy are probably of no use because of the benign nature of this disorder [8, 15].

In conclusion BCM is a rare condition where there is no standardized therapy. However, in light of available data complete resection is a reasonable approach, and in case of recurrence the use of anti-estrogens, gonadotrophin-release hormone analogues, CT-guided aspirations or cisplatin-based chemotherapy are available options [16].

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