Benign Cystic Mesothelioma of the Peritoneum

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Benign cystic mesotheliomas are uncommon mesothelial proliferations that tend to recur but are nevertheless benign lesions. These lesions occur predominantly in women in the reproductive age group (5:1 female/male ratio). The natural history and pathogenesis of this condition remain poorly defined from the limited information available. Most patients have a history of a previous pelvic operation, endometriosis or pelvic inflammatory disease. Many patients are asymptomatic and the tumor is typically found incidentally, but symptoms of variable intensity can occur depending on the size of the tumor. The classic presenting signs and symptoms are abdominal pain, tenderness, and an abdominal or pelvic mass [1,2].

The preoperative diagnosis of benign cystic mesotheliomas is difficult to make. Although diagnostic procedures such as ultrasound, computed tomography and magnetic resonance imaging demonstrate an abnormality suggestive of this disorder, confirmation of the diagnosis is accomplished only at surgery [3]. Histologically, multiple cystic spaces are observed, lined by flat and cuboidal mesothelial cells, and immunohistology shows calretinin and keratin staining. We report the case of a young male with benign cystic mesotheliomas and discuss the diagnostic workup and treatment of this rare disease.

Patient Description

A 23 year old previously healthy man complained of progressive abdominal pain, intermittent abdominal bloating and weight loss during the course of 2 years. Physical examination revealed a mass in the left upper quadrant of the abdomen. Complete blood count, erythrocyte sedimentation rate, liver and renal function tests were within normal limits. A CT scan of the abdomen showed a well-defined, non-calcified multilocular cystic mass, 17 x 7 cm in the left upper quadrant close to the spleen, with displacement of the small bowel. Ultrasonography showed a cystic mass with poorly defined septation, separated from the spleen and tail of the pancreas. Colonoscopy was unremarkable. Carcinoembryonic antigen and CA19-9 tumor markers were within normal limits.

Subsequent diagnostic laparoscopy revealed a conglomerate of cystic structures ranging in size from 1 mm to 3 cm in diameter in the greater omentum. In addition, the greater omentum, and the parietal and visceral peritoneum contained diffused small nodules approximately 1 cm in diameter with clear serous fluid. Many cysts were localized around the appendix. No cystic lesion was noted in the liver, kidney or spleen. Frozen-section laparoscopic biopsies from the cystic mass were taken. Since the results of the frozen sections were non-conclusive, laparotomy was performed and the omental mass as well as the appendix were removed. The postoperative course was uneventful. The patient remains free of symptoms after 12 months of follow-up.

Pathologic findings

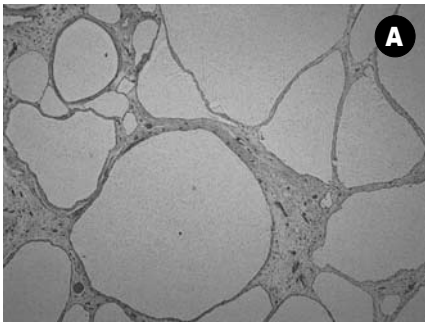
The specimen consisted of two omental pieces measuring 17 x 7 x 5 cm and 16 x 10 x 0.4 cm, with similar appearance. The omentum showed multicystic lesions with a sponge-like appearance, and cysts 0.2–5 cm in diameter with a thin translucent wall and slightly mucinous clear content. Between the cysts fibrous tissue was present. Microscopic examination demonstrated multiple cystic spaces lined by flat to low cuboidal cells, with occasional hobnail cell features, and no atypia or mitoses. The cysts were separated by fibrous septa with areas of acute and chronic inflammation and slight stromal cell proliferation [Figure A]. The cells lining the cysts stained positive for cytokeratin and calretinin markers on immunohistochemical stains [Figure B]. A diagnosis of benign multicystic mesothelioma was made. The appendix was unremarkable.

Comment

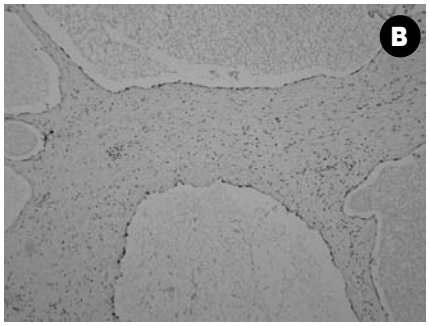
Benign cystic mesotheliomas are borderline lesions because they do not metastasize; however, they have a high propensity for local recurrence. The clinical features of this entity as well as intraoperative findings and pathologic characteristics were clearly explained in a recently published review and isolated case reports [1-3].

The pathologic analysis is of particular importance since it allows differential diagnosis of these lesions from other clinically undistinguishable mesenteric tumors [1,3]. Yet, a number of issues remain to be clarified. First, preoperative diagnosis is still problematic. Ultrasound and CT do not differentiate benign cystic mesotheliomas from other cystic lesions. While fine-needle aspiration could be theoretically exploited as a diagnostic tool, in most cases this method is not informative. MRI reveals a multiloculated cystic mass showing hypointensity on T1-weighted images and hyper or intermediate intensity on T2-weighed images and, therefore, may be very helpful for making a preoperative diagnosis. Laparoscopy is the most accurate diagnostic method since it allows local biopsy of the suspected tissue specimens, however it is an invasive procedure [1,2]. Second, the literature does not provide uniform treatment strategies for peritoneal cystic mesothelioma. Complete eradication of all peritoneal cysts (peritonectomy) is the mainstay of therapy. Interestingly, the lateral margins of the lesion can often be seen to merge with the surrounding normal mesothelial covering, making complete excision difficult if not impossible.

Tumor recurrence is relatively common (50–60%). Given that there are not more than 140 reported cases of benign multicystic mesothelioma in the literature, it is unrealistic to plan randomized clinical trials to evaluate whether aggressive therapy as compared to peritonectomy alone will result in survival benefit. There are several reports on other therapeutic approaches. Cytoreductive surgery with heated intraoperative intraperitoneal chemotherapy using cisplatin or doxorubicin has been effective in several series of patients with peritoneal dissemination, demonstrating a prolonged disease-free survival [3].



[A] Numerous mesothelial lined cysts embedded in a delicate fibrous stroma (hematoxylin & eosin).



[B] The lining cells are positive for calretinin on immunohistochemical stain.

Based on the predominance of benign multicystic mesothelioma in women in the reproductive age, three case reports suggest some degree of hormonal involvement of the tumor. The use of tamoxifen and long-acting GnRH analogues leads to a reduction in cyst volume and cyst growth. Although immunohistochemical detection of female sex hormone receptors in benign multicystic mesothelioma is uncommon, the focal presence of estrogen receptors and/or progesterone receptors in some lesions does suggest hormonal manipulation as a therapeutic option for tumor reduction before anticipated surgery or management of recurrent lesions in some cases [1,4]. Rosen and Sutton [5] used potassium titanyl phosphate laser to treat these lesions. The laser penetrates to a depth of 2 mm, its primary effect being coagulation and vaporization. The authors consider this method to be efficacious in the treatment of multiple sites disease and, in addition, may reduce the need for repeated laparotomies [5]. It should be emphasized that these are experimental treatments and evaluating their results is difficult because of the rarity of the disease. Thirdly, there are two case reports of a benign to malignant transformation of peritoneal cystic mesothelioma [2]. This fact mandates systematic clinical follow-up of these patients for prolonged periods, perhaps for life. Unfortunately, difficulty in further follow-up is compounded by the fact that there are no reliable clinical or imaging features, or tumor markers, pathognomonic for diagnosis. The expected finding of multiple intraabdominal cystic lesions is rarely seen on CT or MRI.

In conclusion, we present a rare case of a young male with benign cystic mesothelioma that was treated solely by surgery. Because of the rarity of this entity we could not find in the literature the optimal therapeutic management for our patient. Although there are no efficient tools for early diagnosis, a long-term follow-up is needed due to the high incidence of recurrence.

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