Case report

Benign cystic mesothelioma: A rare cause of ascites in a case with familial Mediterranean fever

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Received on February 21, 2003; accepted on June 4, 2003.

Clin Exp Rheumatol 2003; 21 (Suppl. 30): S41-S43.

Key words: Ascites, benign cystic mesothelioma, familial Mediterranean fever.

ABSTRACT

Benign cystic mesothelioma (BCM) is a rare neoplasm of the peritoneum, consisting of solitary or multiple cysts arising from mesothelial cells. Here we report a patient with a previous diagnosis of familial Mediterranean fever (FMF) presenting with abdominal distension and ascites which were found to be due to BCM. The co-existence of these two entities has not been reported previously. Ascites as the presenting feature of BMC is also a rare observation.

Introduction

Benign cystic mesothelioma (BCM) is a rare tumor of the abdominal and pelvic peritoneum consisting of solitary or multiple cysts arising from mesothelial cells, and was first described in 1979 (1). It presents with abdominal pain, distention and other mass symptoms which are due to compressive effects. Although diagnostic modalities such as ultrasonography, computed tomography or magnetic resonance scan can suggest the diagnosis, confirmation can be had only at surgery (2, 3). Management consists of surgical excision, which unfortunately is not always curative since recurrences have often been described (4).

Familial Mediterranean fever (FMF) is an inherited disorder characterized by episodes of fever, and abdominal, chest and/or joint inflammation. Recurrent peritoneal inflammation may cause peritoneal fibrosis and adhesions, but ascites is a rare finding (5).

Although the association between malignant mesothelioma and FMF has been reported in previous reports (6-9), the co-existence of FMF and benign cystic mesothelioma has not been described. We report a patient known to have FMF for 14 years, who presented with ascites that was shown to be secondary to BCM.

Case report

In October 2001, a 25-year-old white female patient was admitted to our hospital because of abdominal distention. In 1988 she had been diagnosed with FMF because of recurrent fever and abdominal pain attacks for two years. She was administered 1.5 mg/day of colchicine and responded with total remission. Five years earlier she was seen in a gynecology department because of infertility. During the work-up moderate ascites was detected, but she was lost to follow-up.

On admission, her abdomen was distended due to ascites. Her physical examination was otherwise normal. Routine laboratory tests including blood biochemistry, erythrocyte sedimentation rate, C-reactive protein,WBC, Hb, platelet count and urine analysis were within normal limits. Macroscopically the ascitic fluid was yellowish and clear, and the biochemical examination revealed, albumin 3.32 g/dl, and LDH 185 U/L (serum albumin: 4.2 g/dl, LDH: 224 U/L). Her serum-ascites albumin gradient, which was below 1.1, confirmed exudative ascites. Acid-resistant bacilli were not seen on microscopic examination and the culture of ascites was sterile for aerobic and anaerobic microorganisms and tuberculosis. There were no tumor cells on pathological evaluation. On genetic analysis of the MEFV gene, she was found to be M694V homozygous.

Abdominal ultrasonography and computerized tomography showed moderate ascites. Magnetic resonance imaging revealed multiple thin-walled cysts (Fig. 1). For further evaluation a diagnostic laparoscopy was performed. Laparoscopy revealed numerous multiple thin-walled cysts involving the peritoneum, spleen, liver, pancreas and pelvic viscera, with dimensions ranging from 1-4 cm and containing clear, serous fluid. Only one cyst was excised

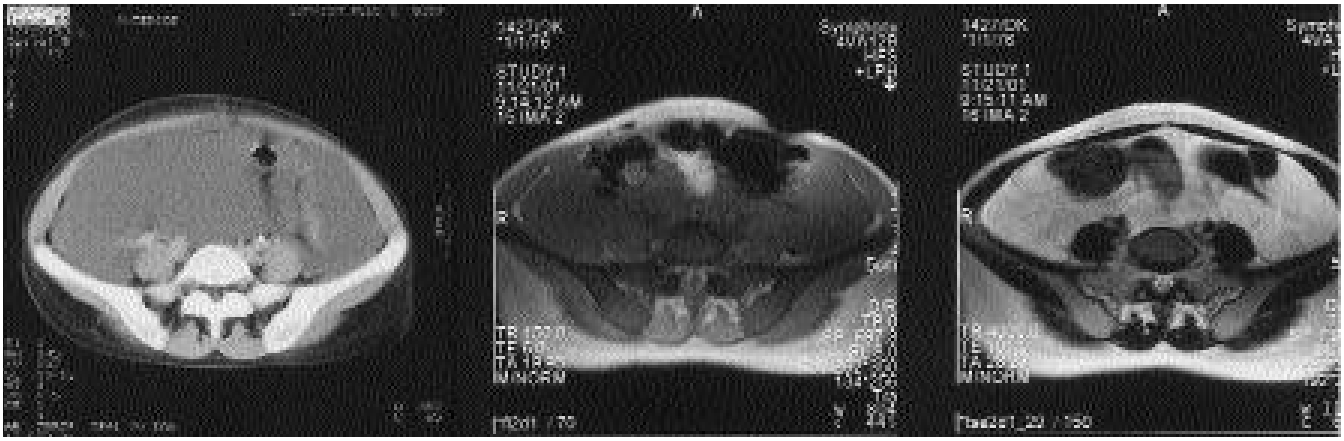

Fig. 1. Cross-sections of images obtained by (**a**) computerized tomography (CT), (**b**) axial T1A magnetic resonance imaging, and (**c**) axial T2A magnetic resonance imaging crossing the same plane. The cysts observed during the operation cannot be differentiated from ascites in the CT. Only a few can be perceived on the T2 cross-sections of MRI.

for diagnosis. The wall contained a smooth lining and a considerable amount of fibrous tissue. The content was yellow and gelatinous. On microscopic examination, the cyst was typically lined with a single layer of flat to cuboidal or hobnail-shaped cells which formed small papillae in some parts of the cyst. The stroma consisted of loose fibrous tissue with a sparse chronic inflammatory cell infiltrates (Fig. 2). In the immunohistochemical study, the lining cells were stained strongly positive with cytokeratin and negative with CD31. These findings were in favor of a mesothelial, rather than endothelial origin.

Discussion

BCM occurs predominantly in women of reproductive age and tends to recur locally but has no malignant potential (10). Clinically it may be asymptomatic or present as abdominal discomfort, an abdominal or pelvic mass, and pain. Although its etiology remains unclear, infection, foreign bodies, chronic peritoneal irritation and endometriosis have been hypothesized as risk factors (11).

Our patient was of child-bearing age, although she was infertile. Her hormonal levels were within normal limits, but on laparoscopy there were peritoneal adhesions which could explain her infertility. This does not contradict with the previous literature which suggests that this tumor is associated with hormonal sensitivity (12).

The co-existence of FMF with malignant peritoneal mesothelioma and with pulmonary mesothelioma have been described in three cases (6-8) and in 5 cases (9) respectively. Recurrent serosal inflammation observed in FMF has been suggested to play a role in the pathogenesis of this malignancy. However, in almost all cases of FMF, peritoneal attacks cause only limited peritoneal fibrosis or, less commonly, encapsulating peritonitis (7).

The co-existence of BCM and FMF, two different diseases that may cause abdominal pain, has not been reported before. Chronic peritoneal irritation is hypothesized in the etiology of BCM (11). Whether the peritoneal attacks of FMF play a role in the development of BCM is not clear, although the full remission obtained with colchicine treatment in this patient makes this explanation unlikely.

Generally patients with BCM complain of abdominal pain and a mass which were absent in our patient. Instead, she presented with ascites which has been reported before only in a child with BCM (5). Profuse and recurrent ascites is unusual in FMF, so the appearance of permanent ascites in FMF patients should alert the physicians to look for underlying causes. The presence of ascites in FMF patients suggests a nephrotic syndrome secondary to amyloidosis; however, massive ascites has been reported in cases with associated malignant peritoneal mesotheliomas (7, 8). The etiology of ascites in malignant peritoneal mesothelioma is due to peritonitis carcinomatosa, but its cause is unclear in BCM.

This case is important in that it represents the first description of benign

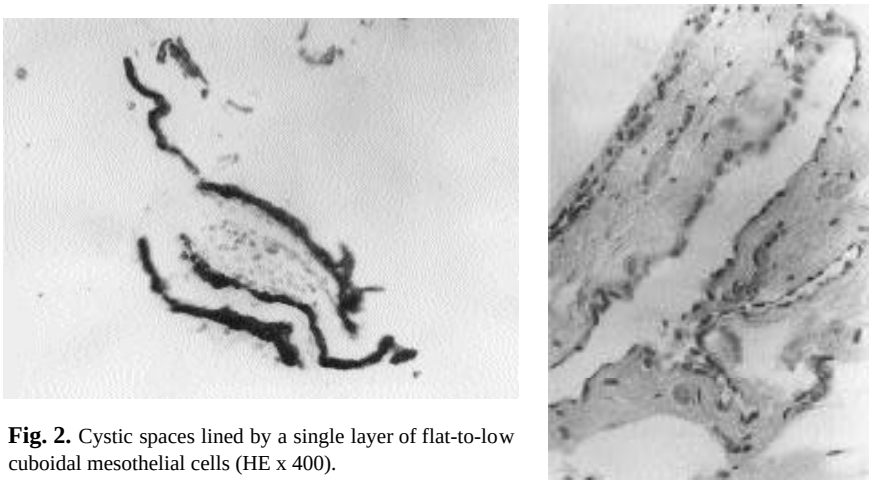

Fig. 2. Cystic spaces lined by a single layer of flat-to-low cuboidal mesothelial cells (HE x 400).

multicystic mesothelial proliferation in an FMF patient. In addition, it is the first case of adult BCM presenting with ascites.

Acknowledgements

We wish to acknowledge Prof. Dr. Gülen Dogusoy for describing the histopathological features; Prof.Dr. Adnan Yaldiran, Prof. Dr. Fikret Sipahioglu and Prof. Dr. Esin Öztürk for their valuable help in evaluating the patient; and Prof. Dr. Huri Özdogan for helpful suggestions and valuable comments during the preparation of the manuscript.

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