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CASE REPORT

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Giant cystic lesion mimicking pseudomyxoma peritonei

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Abstract A case of intraabdominal giant cystic lesion that raised concern for pseudomyxoma peritonei preoperatively is presented here. A young woman underwent work-up for abdominal distension. Abdominal computed tomography suggested a diagnosis of pseudomyxoma peritonei. However, cystic degeneration of a giant myoma was found at surgery. There was no evidence of malignancy on histopathologic examination. This case illustrates that giant cystic lesions can mimic mucinous adenocarcinoma and pseudomyxoma peritonei.

Keywords Degenerating uterus myoma · Pseudomyxoma peritonei · Diagnosis

Introduction

Pseudomyxoma peritonei, a rare condition characterized by diffuse collections of gelatinous fluid, is associated with mucinous implants on the peritoneal surfaces and omentum. The computed tomography (CT) findings in cases of peritoneal adenomucinosis are pathognomonic when proper triple contrast study is used [1]. Different conditions simulating pseudomyxoma peritonei preoperatively have been reported in the literature [2–5]. Here we describe a case of benign giant cystic lesion in a young woman that was clinically and radiologically confused with mucinous adenocarcinoma on preoperative investigation.

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Case report

A 33-year-old woman was referred to the surgical outpatient clinic with a history of abdominal distention for the previous 3 months. On clinical examination, an abdominal enlargement with no masses was found. She was referred for CT scanning but was lost from follow-up for 6 months because of anxiety over any finding. Half a year later she underwent abdominal CT scanning that revealed a diagnosis of pseudomyxoma peritonei (Fig. 1). Her blood tests were normal. Tumor markers revealed a slow- rising CA125 level (90.4) and normal CEA and CA19-9 (0.60 and 3.19, respectively). Preoperative ultrasonography did not show the gynecological source of the finding. Extrinsic pressure to the cecum and terminal ileum was seen on barium enema. The patient underwent an exploratory laparotomy. Intraoperative findings included a giant cystic lesion measuring 34×30×6 cm and weighing 2,450 g exiting from the uteral body on a short stalk (Fig. 2). Another smaller intrauterine nodule was present. Both fallopian tubes and ovaries were normal. Resection of the cystic lesion and myomectomy were performed. Cystic degeneration of a giant myoma was found on histopathologic examination. The patient was discharged uneventfully on the 7th postoperative day.

Discussion

Pseudomyxoma peritonei is characterized by the gradual accumulation of large volumes of mucinous ascites in the peritoneal cavity. It occurs in two of every 10,000 laparotomies and is two or three times more common in females than in males [6]. In most patients, pseudomyxoma peritonei originates in the appendix and then spreads to involve other sites, including the ovaries [7].

Clinical presentation is nonspecific. Suspected acute appendicitis, increasing abdominal girth, or a new-onset hernia are the most common presenting symptoms [8].

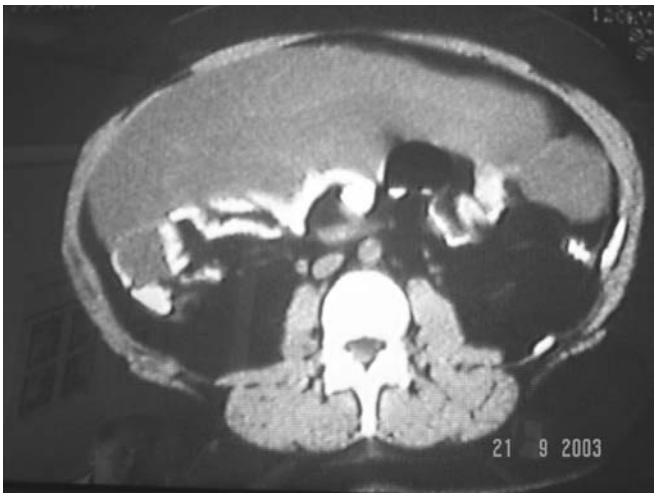

Fig. 1 Preoperative diagnosis of "pseudomyxoma" by CT scan

Abdominal and pelvic CT has been used with great success to characterize the pseudomyxoma peritonei syndrome and differentiate it from peritoneal carcinomatosis. The typical radiological signs of peritoneal adenomucosis on CT scan include relative sparing of the small bowel and mesentery, which is clearly separated from a large volume of mucinous tumor; central displacement of bowel loops; scalloping of the liver and spleen; ascites; and septations [1, 9]. The mucinous material is of fatty density. Curvilinear calcifications and omental thickening are other highly suggestive futures. In the latest studies, parenchymal organ invasion, infiltrative changes in the mesentery, and peritoneal nodules may be present. Magnetic resonance imaging (MRI) has demonstrated the morphological futures shown by CT scanning. In addition, T2-weighted images enable differentiation between mucinous and fluid ascites. However, there have been very few published reports involving MRI scans, and application is limited by cost and availability [10]. Many reports refer to cases discovered before ultrasonography, CT, or MRI were used regularly in investigating abdominal complaints.

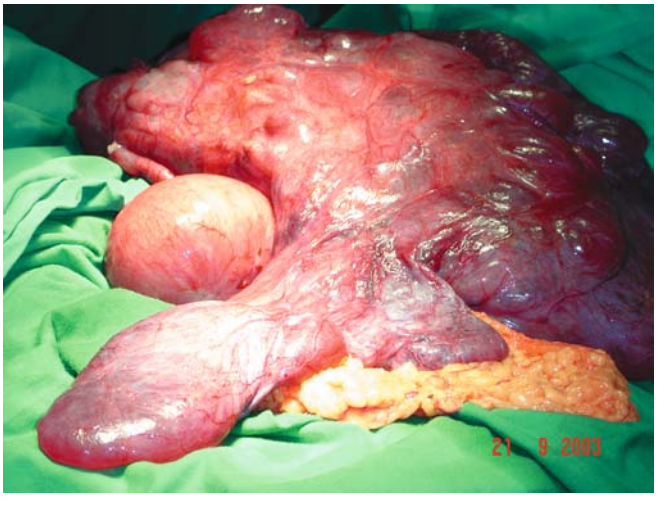

Fig. 2 Intraoperative finding: giant degenerating uteral myoma

Surgical debulking is currently regarded as a primary treatment approach. Sugarbaker advocates that aggressive cytoreductive surgery combined with intraperitoneal chemotherapy improves survival [11].

Various intraabdominal pathologies confused with pseudomyxoma have been reported. Cases of malignant peritoneal mesothelioma [4, 12], abdominal hydatidosis [13], endometriosis with myxoid changes [5], degenerated subserous leiomyoma of the uterus [2], and giant retroperitoneal mucocele [3] suggested a diagnosis of pseudomyxoma peritonei on preoperative CT scan or ultrasonography. However, preoperatively diagnosed intraabdominal echinococcosis [14] and splenic mass [15] were missed with pseudomyxoma intraoperatively. Preoperative diagnosis of giant cystic lesions is difficult as well. Several reports have described missing ovarian cysts with ascites [16, 17].

Malignant peritoneal mesothelioma is an asbestos-related malignancy that usually occurs in males over 50 years old who have a history of asbestos exposure. The CT findings of mesothelioma vary: Approximately 80% of patients have a variable amount of ascites, associated with omental and mesenteric infiltrations, intraabdominal masses (mainly in the upper abdomen), and scattered nodules that tend to envelop viscera and bowel loops [18].

Sugarbaker advocates measuring CEA levels but does not report data on his patients' levels. A few authors have retrospectively reported the levels of CEA and CA19-9 [19]. These data indicate only that levels of CEA, and perhaps CA19-9, are frequently raised in patients with pseudomyxoma peritonei. In a report of four patients with pseudomyxoma peritonei and mucinous ovarian tumors, three also had elevated CA125 levels [20]. In our patient, CEA and CA19-9 levels were normal, but CA125 was slightly increased. These changes were not specific for any diagnosis.

Laparoscopic surgery has been successfully applied to many kinds of gynecological pathology. Currently, giant and benign ovarian cysts can be treated laparoscopically [21]. If the diagnosis is made preoperatively, a minimally invasive approach is preferred for our patients. The value of diagnostic laparoscopy in pseudomyxoma is unknown. A difficult abdominal entrance due to disease process near the abdominal wall and the possibility of bowel injury contraindicate laparoscopy in such cases. However, diagnostic laparoscopy may be useful in cases of unambiguous preoperative diagnosis for investigation of the abdominal cavity and laparoscopic treatment of intraabdominal pathology if possible.

We suggest that preoperative diagnosis of various rare benign and malignant conditions that mimic pseudomyxoma peritonei is difficult. A combined team consisting of the surgeon, gynecologist, radiologist, oncologist, and anesthesiologist should participate in evaluating and treating such patients. Surgeon and

patient should be prepared for major surgery that surprisingly may be changed on the operating table.

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