Case Report

Sarcomatous malignant mesothelioma of the peritoneum: A case report

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Abstract

Malignant mesothelioma of the peritoneum is an uncommon and fatal disease. We report here a case of diffuse sarcomatous malignant mesothelioma of the peritoneum. A 72-year-old woman was admitted to our hospital because of lower abdominal pain. Results of the physical examination suggested massive ascites, and she had no history of any other illness preceding the onset of distention. Computed tomography scan and magnetic resonance imaging confirmed gross ascites. The preoperative diagnosis was peritonitis carcinomatosis probably due to gastrointestinal carcinoma, however endoscopic examinations and fine needle aspiration cytology failed to identify the cause of the condition. The patient was scheduled for diagnostic laparoscopy. A laparoscopic examination revealed extensive intraperitoneal dissemination of a malignant neoplasm without a recognizable primary site. During observation, multiple nodules from several millimeters to 3 cm in diameter were found throughout the peritoneum. Laparoscopic biopsy of the peritoneal nodule was performed. Pathologic studies of the peritoneal nodules showed a malignant methothelioma of the sarcomatous type. One month after laparoscopic examination, the patient died of respiratory failure. Diagnosis of malignant mesothelioma of the peritoneum is generally difficult. Sarcomatous malignant mesothelioma of the peritoneum is considered fatal. Over twenty such cases have been reported in this country.

(Key words: malignant mesothelioma, peritoneum, laparoscopy)

Introduction

Malignant mesothelioma of the peritoneum represents an extremely rare malignancy of the abdominal cavity and accounts for 10–20% of all mesotheliomas1). Most patients present with chronic abdominal pain and ascites which becomes progressive at a relatively late stage of the disease. Abdominal complaints are often vague, and physical examination findings are non-specific, making early exploratory surgery difficult. We report a case of malignant mesothelioma of the peritoneum with sarcomatous histology.

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Case report

The 72-year-old female patient had been experiencing lower abdominal pain and appetite loss over a 1-month period. Except for bronchial asthma, she was in relatively good health. She had no history of asbestos exposure. On admission, the abdomen was slightly distended, and after admission, the right lower quadrant abdominal pain was exacerbated. Pulse was 80 beats per min, blood pressure was 140/87 mmHg, and body temperature was 37.0°C. No lymph node swellings were found. Examination of the heart and lungs was unrevealing. No abdominal mass was palpable. Laboratory findings were: hemoglobin 12.3 g/dl, WBC 6400 μl, and platelets 245,000. The chemistry studies were within normal limits except for CRP of 3.5 mg/dl. Tumor markers were measured; CEA was 2.2 ng/ml, CA19-9 was 15.6 U/ml, and serum CA125 was elevated at 840 U/ml. A chest X-ray showed no abnormalities, particularly no pleural effusion or infiltration. A plain abdominal X-ray revealed several air images in the small intestine. Abdominal ultrasound studies showed marked ascites and a few dilated loops of the small intestine. Computed tomography studies revealed irregularly distributed ascites from the peritoneal cavity to the pelvic cavity, peritoneal and mesenteric thickening, and disseminated nodular lesions involving the omentum and peritoneum from several millimeters to centimeters in diameter (Fig 1). A large tumor about 7 cm in diameter was found in the pelvic cavity, which

![Figure 1](image1.png)  
**Figure 1** Computed tomography of the upper abdomen showing presence of disseminated nodular lesions (arrows).

![Figure 2](image2.png)  
**Figure 2** Magnetic resonance imaging showing the large tumor in the pelvic cavity (arrows).
was not continuous to the uterus. On magnetic resonance imaging, the pelvic tumor showed a high signal on T2-weighted sequences with heterogeneous enhancement (Fig 2). The most probable preoperative diagnosis was a disseminated carcinomatosis due to gastrointestinal carcinoma or ovarian carcinoma. Gastroscopy and colonoscopy did not show any abnormalities. An ultrasound guided fine needle aspiration revealed unremarkable serous fluid, which was slightly bloody, and a result of class II cytology. The bacterial culture of ascitic fluid and Rivalta reaction were negative. The patient was scheduled for diagnostic laparoscopy. Upon entry into the abdomen, diffuse tumor dissemination with mucinous accumulations was observed. The omentum presented with numerous papillary nodules measuring from several millimeters to 3 cm in diameter. Subdiaphragmatic spaces, liver, pancreas, stomach, and colon had no evidence of a recognizable primary malignant tumor. The pelvic tumor involving the peritoneum and omentum was present, but tumor invasion of either the ovary or uterus was not confirmed. Biopsy specimens were obtained from the pelvic tumor. Histologically, biopsy specimens showed extensive proliferation of tumor cells (Fig 3). In some areas, the tumor cells were arranged in sheet like structures, while scattered tumor cells and tumor cells arranged in papillary structure were seen in other areas. Histological studies suggested the diagnosis of diffuse malignant mesothelioma of the sarcomatous type. Immunohistochemistry was performed to confirm the diagnosis of malignant mesothelioma. The tumor cells demonstrated

Figure 3  The bunched tumor cell shaped spindles point in various directions (H&E, ×100).

Figure 4  The tumor shows strong cytoplasmic reactivity for vimentin (H&E, ×100).
positive staining for cytokeratin (AE1/AE3, CAM5.2) and vimentin, but lacked immunoreactivity for Ber-Ep4, calretinin, and EMA (epithelial membrane antigen) (Fig 4, 5). CA125 in ascitic fluid was elevated at 3400 U/ml.

The patient made an uneventful postoperative recovery. Two weeks after laparoscopic examination, the patient was treated with TS-1, but abdominal distention was exacerbated and abdominal X-ray showed several air-fluid levels in the small intestines. The patient’s appetite decreased gradually. Three weeks later, right pleural effusion was recognized by chest radiograph. Four weeks later, the patient died of respiratory failure. Autopsy was not performed.

Discussion

Malignant mesothelioma of the peritoneum is usually (about 85% of cases) diffuse and widespread at presentation. Localized malignant mesothelioma is rare (about 15% of cases), and includes adenomatoid, papillary, cystic and multicystic types. In the Annual of the Pathological Autopsy Cases in Japan, 1,213 cases of pleural mesothelioma (68.0%), 431 peritoneal cases (24.1%), 108 pericardial cases (6.1%), six tunica vaginalis testis cases (0.3%), and 28 “others” (1.6%) were reported. The most common presenting complaints are increased abdominal girth and pain\(^5\). A high level of serum CA125 was observed in our case. An association between malignant mesothelioma of the peritoneum and high serum levels of CA125 has been reported. CA125 was originally used as a marker of Mullerian differentiation and is expressed by a number of nonmucinous ovarian neoplasms including papillary forms. It has also been reported in normal peritoneal mesothelium. An association between peritoneal mesothelioma and high serum levels of CA125 has been reported in several cases\(^9\). Asbestos exposure is a known risk factor for malignant mesothelioma and is associated with pleural mesothelioma, while the correlation between asbestos and peritoneal mesothelioma is less clear. It is not clear how inhaled asbestos induces peritoneal neoplasms, but it has been reported that asbestos fibers can reach areas in the peritoneal cavity where some mesotheliomas develop. Sugarbaker reported that 20 of 51 peritoneal mesothelioma patients had a history of exposure to asbestos\(^4\). Our case had no history of asbestos exposure. Bani-Hani et al. reported that there
was no relationship between asbestos exposure and peritoneal mesothelioma among women. Computed tomography is highly variable, but rarely establishes or excludes definitive diagnoses since differentiating peritoneal mesothelioma from other intra-abdominal malignancies is very difficult. Bani-Hani et al. reported that none of the seven patients with peritoneal mesothelioma was diagnosed preoperatively. The overall sensitivity of cytologic examination for the diagnosis of malignant pleural mesothelioma was 32%. On the other hand, Kitahara K et al. reported that the positive of cytologic examination of malignant mesothelioma of the peritoneum was 12.5%. Sugarbaker et al. reported that definitive diagnosis was not achieved by fluid sampling in any of 68 patients with peritoneal mesothelioma. Furthermore, multiple laparoscopic biopsies of peritoneal lesions enabled histological diagnosis.

Malignant mesothelioma of the peritoneum is diffuse due to the nature of the mesothelial linings of peritoneal cavities. Advanced-stage tumors may invade locally and superficially into adjacent structures, and metastasis to retroperitoneal and pelvic lymph nodes is not uncommon. In the later stages of the disease, plaques and large masses develop as described in the current patient. In typical cases of malignant mesothelioma of the peritoneum, the parietal and visceral peritonei are diffusely coated with a whitish nodular material. The majority of cases are the epithelial diffuse malignant type in which the mesothelial cells are arranged in tubulopapillary or trabecular formations. The malignant mesotheliomas are sub-divided pathologically into the epithelial (50-75%), sarcomatous (15-20%), or mixed type (20-30%). The sarcomatous type reportedly accounts for 5-13% of all malignant mesotheliomas in this country, and is considered comparatively rare and the most fatal type of mesothelioma. More than 20 cases of malignant sarcomatous type peritoneal mesothelioma have been reported in this country. The median survival time of the sarcomatous/biphasic type was 13 months and survival of the epithelial/papillary type was 55 months, respectively.

In addition, although a number of malignant mesothelioma cases have been associated with an elevated content of hyaluronic acid in pleural or ascitic fluid, a significantly lower concentration of hyaluronic acid has been observed in the sarcomatous type. Most mesotheliomas generally show immunoreactivity for cytokeratin, vimentin and EMA, and no immunoreactivity for Ber-Ep4. Specifically, cytokeratin is expected to stain about 100% of cases, vimentin about 40%, EMA about 80-100%, and Ber-Ep4 about 0-11%. Calretinin immunoreactivity has been identified as a sensitive immunohistochemical marker for mesothelioma. Our tumor showed no immunoreactivity for calretinin, and indeed sarcomatous malignant mesotheliomas reportedly have lower immunoreactivity for calretinin and other markers. Our case appears to be compatible with malignant mesothelioma because vimentin was positive and Ber-Ep4 was negative immunohistochemically.

The prognosis of malignant mesothelioma of the peritoneum is poor, with an average survival length of 1 year after diagnosis. Malignant mesothelioma of the peritoneum has been treated most commonly with various chemotherapeutic such as doxorubicin, cisplatin or cyclofosfamide, but there is no evidence that chemotherapy prolongs survival. Eltabakh et al. reported that cytoreductive surgery and chemotherapy (paclitaxel and cisplatin, in particular) might benefit women with malignant mesothelioma of the peritoneum. Muensterer et al.
reported that cytoreductive surgery and perioperative intraperitoneal chemotherapy successfully treated the intra-abdominal lesions and ascites except for seeding in the port site. Stamat et al. reported that early diagnosis of malignant mesothelioma by laparoscopic biopsy of peritoneal nodules is important to distinguish malignant mesothelioma from benign or localized mesothelioma, so that appropriate treatment can be started promptly.

In conclusion, we experienced a case of sarcomatous malignant mesothelioma of the peritoneum, which was diagnosed by laparoscopic biopsy. Early exploration by laparoscopy is of great diagnostic value when malignant mesothelioma of the peritoneum is suspected. Current treatments for malignant mesothelioma are largely ineffective, and thus novel therapeutic regimens are needed.

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References


肉腫型悪性腹膜中皮腫の1例

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要 約

悪性腹膜中皮腫はまれな疾患であり、予後不良とされる。我々は肉腫型悪性腹膜中皮腫の1例を経験したので報告する。症例は72歳女性、下腹部痛のため入院、発症前に既往疾患はなく、身体所見では腹水が示唆された。CTとMRIで腹水を認め、術前診断としては消化器癌による悪性腹膜炎を疑ったが、内視鏡検査では原発巣となるものは認められず、腹水細胞診でも診断に至らなかった。腹腔鏡検査が行われ、原発不明な進行した腹膜播種を認めた。腹膜全体に数ミリから3センチ大の結節が多数存在し、結節の一部を採取し終了した。腹膜結節の病理診断は、肉腫型の悪性腹膜中皮腫であった。腹腔鏡から4週間後に患者は呼吸不全で死亡した。本症例のように悪性腹膜中皮腫は診断に難渋することが多い。また、肉腫型の悪性腹膜中皮腫は予後不良とされており、我々では2例を超える報告がなされている。

（キーワード：悪性中皮腫、腹膜、腹腔鏡）

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