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Pseudomyxoma peritonei due to adenocarcinoma of the lung: Case report

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Abstract: A rare case of pseudomyxoma peritonei whose primary site was presumed to be the lung is reported. A 76-year-old woman was admitted to Hospital presenting with progressive abdominal distention. She had been admitted twice, 2 and 1 year previously for the evaluation of high plasma carcinoembryonic antigen (CEA) level, of 11.6 ng/ml. Chest computed tomography (CT) scan and chest X-ray film on the third admission revealed a nodular lesion in the left lower lung field, and transbronchial lung biopsy (TBLB) revealed mucus-producing tall columnar epithelial carcinoma. Paracentesis revealed gelatinous ascitic fluid. At laparotomy, appendix and ovary were normal, and there were many small cystic tumors on the peritoneal surface and omentum. The patient died 2 years later, after repeated episodes of dynamic ileus. The lung and abdominal tumors gradually increased in size during the 2-year period, but she developed no respiratory symptoms. Based on both the clinical and pathophysiological findings, the final diagnosis made was pseudomyxoma peritonei whose origin was a lung adenocarcinoma.

Key Words: pseudomyxoma peritionei, lung, adenocarcinoma, CEA

Introduction

Pseudomyxoma peritonei is a clinical entity in which diffuse gelatinous mucinous implants occur on the peritoneal surfaces and omentum. Gelatinous ascites is often massive.[1–4] The majority of cases appear to be complications of primary ovarian and appendiceal benign and low-grade malignant epithelial tumors that have invaded the peritoneum.[4] The following case represents the first, to our knowledge, in which pseudomyxoma peritonei was diagnosed to be of lung origin.

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Case report

A 76-year-old woman was admitted to Tokyo University Hospital on May 28, 1986, the major complaints being progressive abdominal distension and intermittent colicky abdominal pain. The patient had been in this hospital twice previously. Two years prior to this admission (September 12, 1984), she had been admitted because of an elevated carcinoembryonic antigen (CEA) level, of 11.6 ng/ml (normal range <2.5 ng/ml) and tarry stools. Endoscopic examination at that time revealed benign gastric ulcers at the healing stage. No specific disease which would explain the high CEA level was identified by chest X-ray, barium enema, abdominal ultrasound, abdominal computed tomography (CT) scan, or gynecological examination. One year prior to this admission (August 12, 1985), she was admitted again for the evaluation of the persistent high plasma CEA level (up to 40 ng/ml). An abnormal nodular lesion in the left lower lung field was identified by chest X-ray and chest tomography, as well as by chest CT scan. Abdominal CT scan and abdominal ultrasonic examinations showed no abnormal findings. In the interval between the second discharge and the third admission, no clinical data were available, since the patient did not consult our outpatient clinic.

Laboratory findings on the third admission disclosed a plasma CEA level of 412 ng/ml, and hypoalbuminemia, albumin being 2.4 g/dl. Other tumor markers, alpha-fetoprotein (AFP) and carcinogenic antigen (CA)19-9, were within the normal range. Chest CT

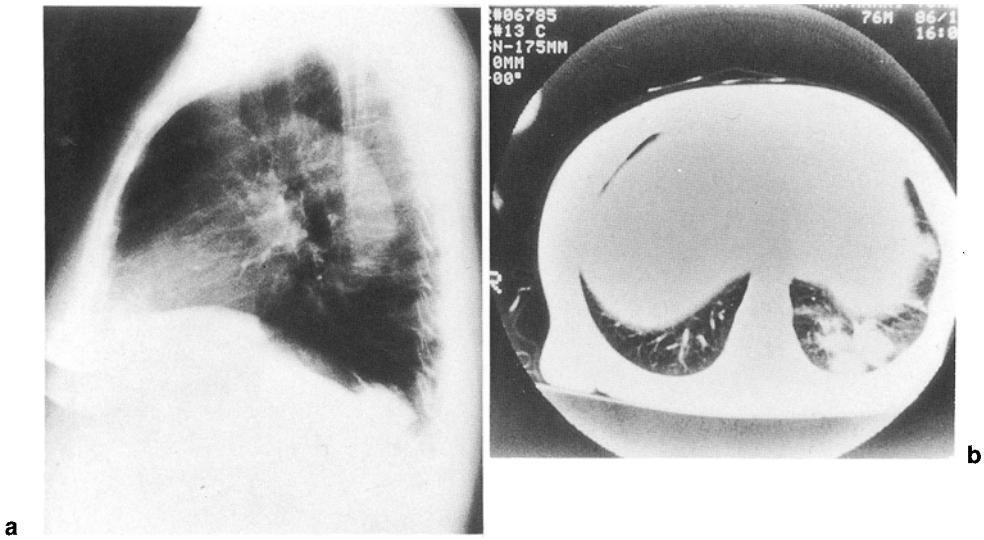

Fig. 1. **a** Chest X-ray film and **b** chest computed tomography (*CT*) scan showing a nodular lesion in the left lower lung field

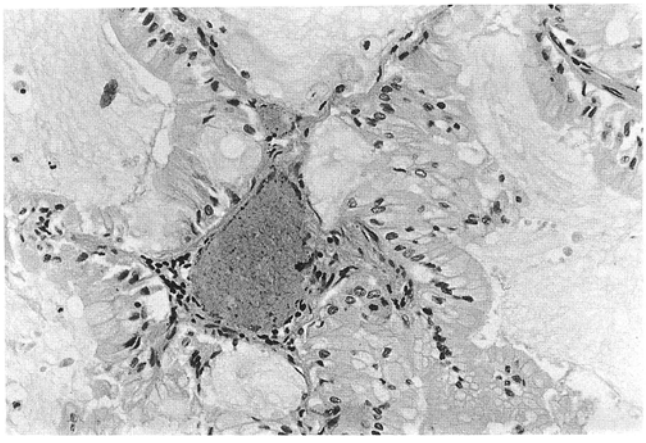

Fig. 2. Histopathological findings of specimen from the nodular lesion in the left lower lung field, obtained by transbronchial lung biopsy, shows adenocarcinoma of the lung (mucus-producing tall columnar epithelial carcinoma). H&E, × 68

scan revealed a nodular lesion in the left lower lung field (Fig. 1). Transbronchial lung biopsy (TBLB) was performed on November 27, 1986, and the histopathology of the biopsied specimen obtained from the nodular lesion was mucus-producing tall columnar epithelial carcinoma (Fig. 2). Abdominal ultrasonic examination suggested multiple abdominal cystic lesions and ascites. Abdominal CT scan showed widely distributed low density areas with septation in both the abdominal and pelvic cavity. Characteristic indentation of the liver surface was also recognized (Fig. 3). Paracentesis revealed yellowish gelatinous ascitic fluid. The diagnosis of pseudomyxoma peritonei was thereby made.

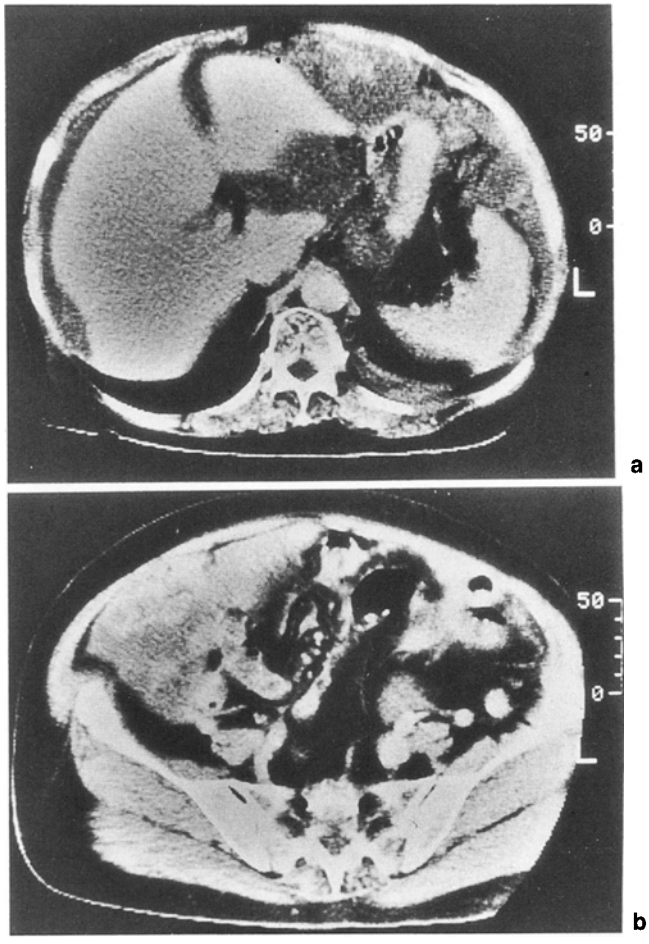

Fig. 3a,b. Abdominal CT scan showing the low density area in the abdominal cavity and pelvic cavity. **a** Section through the upper abdomen reveals ascites and a low attenuated mass producing the characteristic "hepatic scallopping". **b** shows ascitic septation

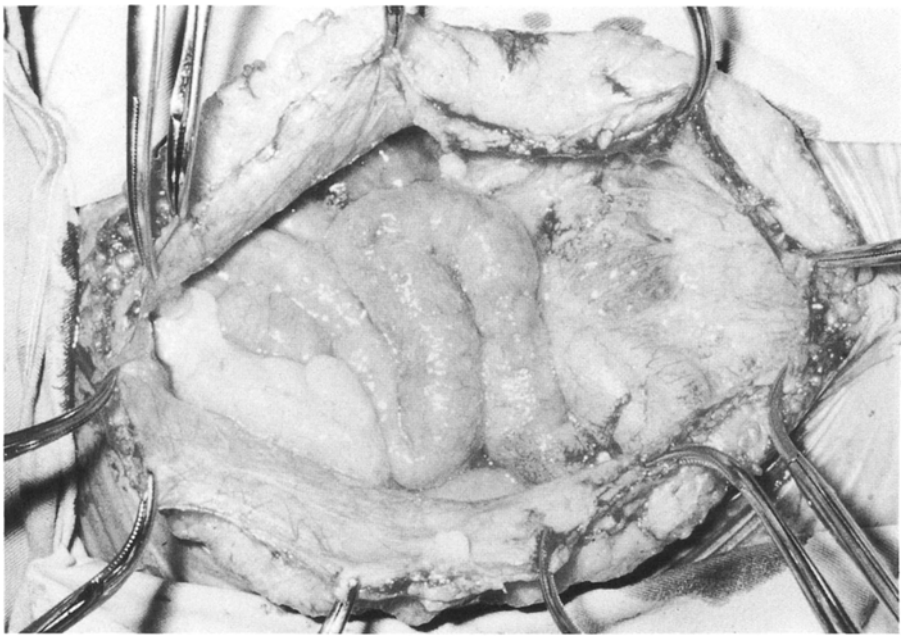

Fig. 4. Operative findings: Laparotomy reveals massive gelatinous substance in the abdominal cavity, and many small tumors attached to the serosa

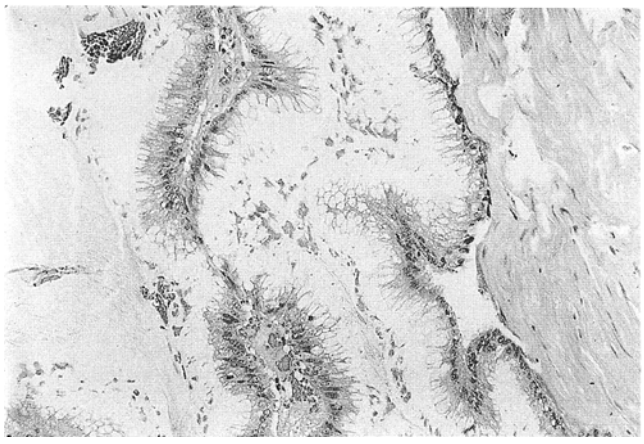

Fig. 5. Histopathological findings of the resected abdominal tumor, showing a mucus-producing metastatic adenocarcinoma. H&E, × 7

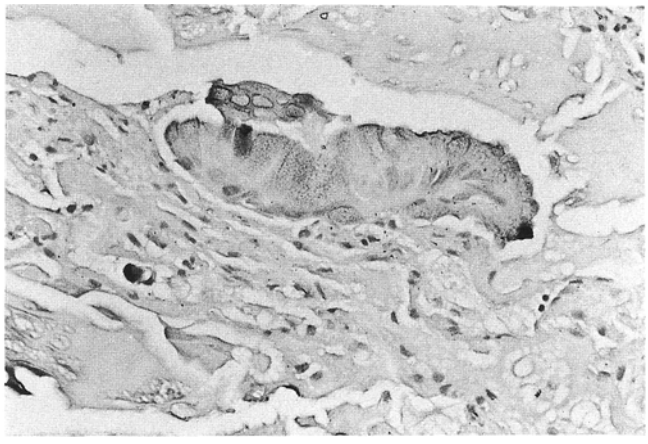

Fig. 6. Mucus staining of the abdominal tumor revealed no difference from that in the lung malignant lesion. This tissue was taken from the abdominal cavity. Alcian blue, × 14

Laparotomy was performed on January 16, 1987, and revealed multiple small cystic tumors on the peritoneal surfaces and omentum (Fig. 4). Nine hundred ml of gelatinous substance was removed from the peritoneal cavity. The histopathological diagnosis of the resected specimen was metastatic mucus-producing adenocarcinoma of the omentum (Fig. 5). No other abnormality was found during the laparotomy. After discharge, the patient was started on chemotherapy with oral 5-fluorouracil, 150 mg per day, and was subsequently followed for 2 years with regular check-ups in the outpatient clinic. The drug had no noticeable effect. A gradual increase in the size of the lung tumor was observed on serial chest X-ray films, but there were no respiratory symptoms. Occasional paracentesis was performed for treatment, both in the outpatient clinic and on readmissions. A high plasma CEA level, up to 500 ng/ml, was maintained, and this fluctuated according to the apparent quantity of the gelatinous ascites (or tumor mass).

In August 1988, the patient was readmitted with suspected ileus. Her general condition gradually deteriorated, with repeated episodes of ileus and gastrointestinal bleeding. She died of renal failure in January 1989. Postmortem examination revealed massive gelatinous ascites in the abdominal cavity, as

well as many abdominal tumor implants and the left lung carcinoma. On examination of the appendix and ovary no specific evidence was found that would suggest either as the primary site of origin of the pseudomyxoma peritonei. The histopathological characteristics of the abdominal tumor appeared to be the same as the those of the lung lesion. Mucus staining was performed on the specimens, and the abdominal tumor and the tissue obtained by TBLB were found to be identical (Fig. 6). Based on both the histopathological and the clinical findings, including the gradual time course, the final diagnosis was established as pseudomyxoma peritonei whose origin was adenocarcinoma of the lung.

Discussion

Pseudomyxoma peritonei is a rare clinical entity. In patients with this disorder, mucinous implants may be found throughout the peritoneal surfaces and omentum. These implants often cause massive gelatinous ascites that sometimes requires repeated paracenteses or laparotomy for treatment.[5] In most cases, pseudomyxoma peritonei is associated with primary appendiceal or ovarian neoplasms with abundant mucin production. Although the malignant nature of the epithelial cells is controversial,[6] the clinical course is characterized by long survival and the absence of extraperitoneal spread.[7] Infrequent origins of pseudomyxoma peritonei have been reported: colon,[8] urachus,[9] pancreas,[10] stomach,[11] uterine corpus,[12] gallbladder, common bile duct, prostata, omentum, etc.[13] The case reported here shares all the typical clinical findings, except for its origin and the extremely high plasma CEA level.

The points of this case can be summarized as follows: First, high CEA was observed. Then an abnormal lung shadow was recognized and TBLB revealed malignant adenocarcinoma. Third, ascites appeared and laparotomy confirmed gelatinous ascites and widespread peritoneal implants. Fourth, there was no difference between the histopathological findings in tissues from the lung lesion and the abdominal tumor.

The slow progression of both pseudomyxoma peritonei and lung adenocarcinoma in this case seems to indicate a common primary site of origin, rather than a double cancer. Reports of extraperitoneal metastasis of pseudomyxoma peritonei are rare.[5,8,14] Sugarbaker and colleagues[8] described two cases of pseudomyxoma peritonei, due to tumors of the appendix and right colon, where implants on the pleural surface were observed without parenchymal invasion. Mets and colleagues[14] reported a case of pseudomyxoma peritonei of appendicular origin where dissemination occurred to the right pleural cavity and to the pericardium, although the pulmonary parenchyma was not involved. Mets et al.[14] also cited several references that described extraperitoneal metastasis and noted that "the invasion of the spleen suggests a highly malignant primary tumor and makes hematogenous dissemination possible; however, since the pleural and pericardial metastasis were superficial, a spread through a diaphragmatic defect seems likely." This speculation might apply to our case also.

So far as the authors know, this is the first case of pseudomyxoma peritonei whose origin was diagnosed as a primary pulmonary neoplasm. Regarding the elevated plasma CEA level, this feature has already been reported.[15,16] The parallel fluctuation in plasma CEA and changes in the amount of ascites (or degree of abdominal distension) suggest that the CEA antibody reacts with the carbohydrate of the mucoprotein present in the ascites. The findings suggest the value of plasma CEA levels in monitoring the progression of the disease condition. Our radiological findings such as the concave depression of the liver margin on abdominal CT scan (so-called scalloping of the liver margin) and the diffuse low density areas with septation in the abdominal cavity (so-called septated ascites), were also compatible with the descriptions in the literature,[17,18] and are well known signs of this rare clinical disorder.

In conclusion, a rare case of pseudomyxoma peritonei whose origin was established as lung adenocarcinoma is reported. Otherwise, the clinical features were as previously described in the literature. A high plasma CEA, up to 500 ng/ml, was observed, and this fluctuated in parallel with the apparent quantity of the gelatinous ascites, suggesting that the mucoprotein present in the ascites reacted with the CEA antibody.

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