Pseudomyxoma of the pleural and peritoneal cavities

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Abstract

Pseudomyxoma peritonei is a rare clinical manifestation of mucin producing adenocarcinomas. An extensively metastased adenocarcinoma developed a pseudomyxoma that affected not only the peritoneal cavity but also the pleura.

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Pseudomyxoma peritonei is a rare clinical entity characterised by mucinous tumour masses in the peritoneum and omentum with gelatinous ascites. It usually originates from adenocarcinoma of the appendix or ovary and less frequently from adenocarcinoma of other organs.1-4

We present a patient with pseudomyxoma that affected not only the peritoneum but also the pleura.

Case report

A 41 year old farmer, a non-smoker, was admitted to hospital in December 1988 because of mild chest pain and breathlessness, clinical signs of ascites, and homogeneous shadowing in the left hemithorax on the chest radiograph (fig 1). His general condition was good. All the results of routine laboratory investigations were within normal limits.

Two and a half years earlier he had had an acute perforation of the appendix, and he had an appendicectomy at a regional hospital. The wound discharged for two months. Six months later he noticed abdominal swelling.

Abdominal ultrasound during his first admission showed massive, loculated ascites without enlargement or metastatic involvement of abdominal organs. Thoracic computed tomography showed thickened pleura and a homogeneous mass in the left hemithorax, described as "a dense pleural effusion" (fig 2).

Abdominal computed tomography suggested "mucinous ascites", partly localised around the liver, following the intestinal curve and concentrated in the pelvis.

Pleural aspiration on several occasions disclosed no malignant cells but a few were found in one sample of pleural gelatinous material. A diagnosis of adenocarcinoma was confirmed by percutaneous biopsy of the main thoracic mass.

Management of bleomycin lung toxicity is frequently difficult, though steroids are widely recommended and evidence supporting their role comes from both animal studies and clinical reports on a few patients.5

Less settled, however, is the value of these agents in advanced bleomycin lung toxicity. Samuels and coworkers7 described a series of five such patients, all of whom received prednisolone in doses of 60-100 mg daily. All five died of acute respiratory failure despite this treatment. Gilson and Sahns6 reported a patient with bleomycin lung toxicity who developed the adult respiratory distress syndrome after surgery and ultimately responded to a combination of antibiotics and methylprednisolone 500 mg a day. Recently Hartmann and colleagues8 also described a patient with life threatening bleomycin lung toxicity successfully treated with methylprednisolone 1 g daily. Our case, taken in conjunction with these, suggests that severe bleomycin lung toxicity may be largely reversible provided that the treatment regimen incorporates corticosteroids in very high doses that are tapered gradually. We elected to use azathioprine in addition because of its immunosuppressive and known steroid sparing effects.

We believe that all patients with bleomycin lung toxicity should receive a trial of corticosteroids. The dose used should depend on the severity of the pneumonitis.

Bronchoscopy showed no abnormality.

Owing to spread of adenocarcinoma within and beyond the abdomen, surgical treatment was not indicated and he was treated with three courses of doxorubicin (Adriamycin), vincristine, cyclophosphamide, and 5-fluorouracil, but these had little effect. Thereafter the patient remained in good general condition for several months, during which he was working, though he tired more easily after hard physical work.

From August 1989 his condition deteriorated, with increasing abdominal distension, umbilical and inguinal hernias, and progressive opacification in the left hemithorax with displacement of the mediastinum to the right. At that time the patient received further cytotoxic chemotherapy, corticosteroids, and intracavitary hyaluronidase (2000 IU) without any effect. Abdominal paracentesis evacuated only 500 ml of a gelatinous substance. Finally he developed oedema of the lower limbs and eventually died in May 1991. Necropsy was not performed.

Discussion

Pseudomyxoma peritonei was first described in 1894 and there have been reports of a few cases since then.1 2 4 The gelatinous material is believed to be produced by a highly differentiated mucin producing adenocarcinoma of relatively low malignancy. Treatment of the abdominal condition has included the use of various anticancer drug combinations, hyaluronidase, corticosteroids, repeated palliative surgical treatment, postoperative radiotherapy, and simple conservative treatment. The survival of patients from the time of diagnosis is over 50% at five years and one patient has survived 24 years.1 2 4

The primary tumour in this case was probably in the appendix and the pleural lesion was due to metastatic spread. This has not been reported previously.