and may require decompressive surgery.⁶⁷ CT scanning or MRI may reveal the characteristic proliferation of retrosternal tissue, but a biopsy specimen may be required to exclude malignancy. A biopsy specimen was not taken in this case due to the overall poor health of the patient.

As far as we are aware, there are no previous reports of sinus formation occurring in SCCH, either over the affected area or peripherally.

The differential diagnosis of SCCH includes malignancies such as plasmacytoma involving the sternum and metastatic thyroid or oral cavity carcinomas. Non-malignant causes include diffuse idiopathic skeletal hyperostosis, Paget's disease condensing osteitis, and osteomyelitis.

Treatment is usually conservative as no specific therapy has been shown to alter the course of the disease. Following stage II, which may last for many years, the final stage III is characterised by ankylosis of the affected joints and

ossification of the cartilage of the ribs. Limited symptomatic relief may be gained with the use of non-steroidal anti-inflammatory agents and corticosteroids.

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Lymphangitis carcinomatosa complicating primary malignant peritoneal mesothelioma

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Abstract

A patient with malignant peritoneal mesothelioma and a diffuse pulmonary infiltrate is described. Computed tomographic scanning suggested lymphangitis carcinomatosa. This was confirmed on transbronchial biopsy to be due to metastatic mesothelioma.

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Keywords: peritoneal mesothelioma, lymphangitis carcinomatosa, metastasis.

Malignant mesothelioma is a rare tumour with an annual incidence of between 0.7 and 1.5 per million.¹² Increasing incidence has been reported in many countries including the United States and, in particular, Australia.¹³ The disease is strongly linked to asbestos exposure, particularly crocidolite. Approximately 10% of mesotheliomas arise from the peri-

toneum.⁴ Lymphangitis carcinomatosa is characterised histologically by diffuse permeation of tumour cells within pulmonary lymphatics. We are not aware of any previously reported examples of lymphangitis carcinomatosa due to malignant mesothelioma.

Case report

A 52 year old man presented with a two month history of weight loss and upper abdominal pain. There was a history of asbestos exposure 20 years earlier. A chest radiograph, taken at the onset of symptoms, was normal. An abdominal computed tomographic (CT) scan showed a diffuse omental mass. Needle biopsy yielded cytologically abnormal mesothelial cells consistent with mesothelioma.

One month later the patient developed a non-productive cough and exertional dyspnoea. Auscultation of the lung fields revealed bilateral fine basal crackles. The chest radiograph on this occasion showed a widespread reticulonodular pattern with septal thickening. A high resolution CT scan of the thorax revealed diffuse nodular thickening with polygon formation (fig 1). Bronchoscopic examination was normal. Transbronchial biopsy specimens showed abnormal epithelioid malignant cells with mild nuclear pleomorphism and prominent nucleoli. These cells were identical to those obtained from the previous omental needle biopsy. Ultrastructurally they showed long branching microvilli consistent with mesothelioma (fig 2). Cytotoxic chemotherapy with cisplatin and doxorubicin was associated with stable disease for three months. The patient died eight months after presentation from respiratory failure due to progressive disease.

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Figure 1 High resolution computed tomographic scan of the thorax showing diffuse interstitial thickening. Peripheral wedge densities are seen in the apical segment of the left upper lobe.

Figure 2 Electron micrograph of a malignant cell from a transbronchial biopsy specimen with long microvilli (length:diameter >20:1). Inset: branching microvilli.

Discussion

The clinical course of abdominal mesothelioma is usually dominated by direct invasion of adjacent structures by the primary tumour. Metastatic disease complicating peri-

toneal mesothelioma is uncommon, even in necroscopic studies.⁵ In contrast, frequent metastases to lymph nodes, lung, liver, and brain have been reported in patients with pleural mesothelioma.⁶ Miliary pulmonary metastases without lymphangitis have been reported as the initial manifestation of pleural mesothelioma.⁷

Lymphangitis carcinomatosa has been associated with many forms of malignancy and, in particular, carcinomas of breast, stomach, lung, pancreas, and prostate. The radiological features of this case are those classically described in lymphangitis carcinomatosa.⁸

Management of peritoneal malignant mesothelioma remains difficult and survival following diagnosis rarely exceeds one year.⁵ Occasional long term survivors are seen following surgical resection.⁹ Only modest activity has been seen in patients with pleural or peritoneal mesothelioma with cytotoxic chemotherapy. The prognosis of patients with metastatic disease, even confined to regional lymph nodes, is particularly poor.¹⁰

With the increasing incidence of malignant mesothelioma, presentations including metastatic disease will become more clinically important. In patients with lymphangitis carcinomatosa underlying malignant mesothelioma should be considered. Electron microscopic examination of transbronchial biopsy material may allow the diagnosis to be rapidly achieved.

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