Sternocostoclavicular hyperostosis presenting with thoracic sinus formation

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Abstract

Sternocostoclavicular hyperostosis (SCCH) is a condition which is well described in the Japanese literature but is rare in Western Europe. It is characterised by pain and swelling in the upper anterior part of the chest, which tends to be progressive. A patient is described with bilateral chronic discharging sinuses over the anterior ends of the clavicles in whom the diagnosis appeared to be one of SCCH.

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Keywords: sternocostoclavicular hyperostosis, thoracic sinus, pustular psoriasis.

Case report

A 75 year old woman of Ashkenazi Jewish extraction was referred because of increasing shortness of breath. For five years she had suffered from recurrent clavicular problems. Initially this had been swelling, pain, and stiffness around the medial ends of the clavicles.
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Figure 1 Discharging sinuses over the upper chest wall and right venous congestion.

that had progressed to chronic discharging sinuses on the upper anterior chest wall. She had developed pustular psoriasis affecting the soles of the feet and the palms of the hands.

On examination she weighed 67 kg and was 145 cm tall. She was not clubbed or cyanosed. The jugular venous pressure on the right side was elevated and non-pulsatile. The veins over the right breast and upper arm were engorged. On the left side of the neck there were discharging sinuses with deformity of the area (fig 1). The right side had a large fluctuant mass at the head of the sternoclavicular joint and a pocketed firmer mass over the right sternomastoid. A surgical scar from her hemicolectomy for diverticulosis still had a dressing on it two and a half years following surgery.

Investigations at the clinic were as follows: ECG revealed sinus rhythm with evidence of right atrial enlargement (P′ pulmonale). Lung function tests showed a mixed and severe obstructive and restrictive pattern with forced expiratory volume in one second (FEV1) 0.571 (36%), forced vital capacity (FVC) 0.91 (45%), and FEV1/FVC 0.64. Chest radiography showed apparent thickening of the anterior end of the left clavicle with some bone destruction (fig 2). The right clavicle was not clearly seen but a possible opacity was noted in the underlying lung.

Ten ml of pus was aspirated from one of the lesions and sent for urgent staining. This failed to reveal the presence of any acid and alcohol fast bacilli (AAFB) and subsequent culture was negative. Her full blood count revealed haemoglobin of 10.9 g/dl, white cell count of 7.2 x 10³/l (lymphocytes 26%, monocytes 10.3%, granulocytes 63.5%). Her erythrocyte sedimentation rate (ESR) was 46 mm in the first hour. Serum calcium was normal and the alkaline phosphatase was raised at 171 U/l (normal 10-110). Rheumatoid factor and antinuclear factor were negative. The sinuses were swabbed repeatedly and specimens were sent for microbiological examination and culture. The swabs failed to grow any organisms including bacteria, fungi, AAFB, or actinomyces.

A computed tomographic (CT) scan showed bilateral destruction of the clavicles and confirmed the presence of a mass in the right upper chest which extended to the right lobe of the thyroid gland. This appeared to be compressing the superior vena cava but not the trachea. In view of the sterile discharging sinuses, anti-tuberculous treatment with rifampicin and isoniazid was started. After eight weeks of treatment there had been no resolution of symptoms nor of the rate of discharge from the sinuses and the medication was therefore stopped. Nebulised salbutamol, 5 mg four hourly, was given for her airways obstruction with an improvement in her spirometric parameters. A fibroptic bronchosopic examination was carried out which showed a normal endobronchial tree. Two years later her physical signs remain unchanged.

Discussion

Sternocostoclavicular hyperostosis (SCCH) is a condition with an equal sex distribution whose peak incidence occurs in middle age. The original reports were from Japan in 1967, although since then cases have been described in America and Europe (there does not appear to be an increased incidence in Japanese migrants to these areas). The disorder was first reported by Sasaki who described a case of bilateral hyperostosis of the clavicles associated with pustulosis palmaris and plantaris. No single aetiological agent has been defined and culture of bone biopsy specimens of affected material has failed to demonstrate an infective cause. There is debate over the pathogenesis of the disease. Kholer et al proposed that it is an ossifying periostitis which begins within bone and progresses to a generalised hyperostosis, but Fritz et al have argued that it is primarily a rheumatological rather than an orthopaedic condition. The diagnosis is primarily clinical and radiological. The most frequently abnormal laboratory investigations include increased ESR, C-reactive protein and alkaline phosphatase. Radionuclide studies show uptake in the sternoclavicular region and may show extrastrernal involvement.

Radiological features of SCCH are said to be characteristic and have been classified into three stages by Resnick. In the case described here the changes corresponded with stage II, with marked destruction of the sternoclavicular joints, the clavicle, and formation of an ossific mass. Extrinsic compression of the subclavian vein and brachial plexus neuropathies due to the effect of local pressure have been reported.

Figure 2 Chest radiograph showing bony destruction of clavicles.
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.

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.1 Increasing incidence has been reported in many countries including the United States and, in particular, Australia.2 The disease is strongly linked to asbestos exposure, particularly crocidolite. Approximately 10% of mesotheliomas arise from the peritoneum.3 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). Bronchosopic 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.