Pancoast syndrome caused by a high grade B cell lymphoma

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Abstract

The case history is presented of a 20 year old man with Pancoast syndrome caused by a high grade B cell lymphoma.

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The Pancoast syndrome is almost invariably caused by a bronchogenic carcinoma, typically a squamous cell tumour.¹ To our knowledge there has only been one other case report of a malignant lymphoma causing the Pancoast syndrome.²

Pancoast first described the superior sulcus syndrome in 1924.³ His original description included pain in the C8-T2 nerve distribution, Horner's syndrome, destruction of upper ribs, and atrophy of hand muscles.⁴

Since this time the Pancoast syndrome has been reported secondary to many unusual causes including multiple myeloma,⁵ hydatid cysts,⁶ aspergillosis,⁷ metastatic uterine cervical carcinoma,⁸ and lymphomatoid granulomatosis.⁹

Case report

A 20 year old man presented to the casualty department with a three week history of sharp left sided chest pain and gradually increasing weakness of his left arm and hand. The pain was infraclavicular and radiated down the inner aspect of his left arm. He reported difficulty with sleeping because of the intensity of the pain.

A friend had recently noticed that his pupils

were of unequal size, the right being larger than the left. He reported no other neurological symptoms, nor did he admit to shortness of breath, cough, sputum production, or haemoptysis.

There was no past medical or family history of significance, and the only medication he was taking was simple analgesia. He smoked about 20 cigarettes per day and occasionally drank alcohol.

On examination he had a small, left sided ill defined scalene node. There was no evidence of anaemia, cyanosis, or clubbing, and he was afebrile. His pulse rate was 80 beats/min, regular and equal in volume in both arms. There was an apparent harsh aortic ejection systolic murmur which radiated to the carotid areas and the back.

Examination of his chest revealed decreased expansion of the left hemithorax and decreased air entry in the left lung. There was mild wasting of his left pectoralis musculature. Abdominal examination revealed nothing remarkable.

His left pupil was considerably smaller than his right. There was anhidrosis and a partial ptosis on the left side, signs in keeping with Horner's syndrome. There was marked wasting of the interossei, thenar and hypothenar eminences of his left hand, and there was a decrease in power of all muscle groups in this hand (grade 3/5). Light touch and pain sensation were decreased in the C8 and T1 dermatomal distribution in his left hand and arm. The rest of the neurological examination was normal.

Full blood count, peripheral film, and blood urea and electrolytes were all normal on presentation. Chest radiography (fig 1) showed only a subtle abnormality, which was only appreciated on a high quality film. An echocardiogram showed no valvular defects or chamber abnormalities to account for the detected systolic murmur.

Magnetic resonance imaging (MRI) of the patient's brachial plexus and upper thorax was performed and showed extensive superior mediastinal (fig 2A) and left hilar lymphadenopathy (fig 2B). There was no evidence of rib destruction on the chest radiograph or on the MRI scan; a bone scan was not performed. A computed tomographic (CT) scan of the brain and cerebrospinal fluid examination showed no abnormality.

The patient underwent a left anterior mediastinotomy, as it was felt that an adequate and diagnostically reliable sample of tissue could not be obtained from the small ill defined scalene node found on clinical examination. At operation it was noted that the left lung was adherent to the mediastinum, which was solid with tumour and hilar gland involvement. Biopsies of the tumour and a sample of bone marrow were taken.

Histological examination of the samples showed the mass to be a high grade non-Hodgkin's B cell lymphoma of centroblastic, pleomorphic subtype. The bone marrow specimen showed no evidence of infiltration by lymphoma or other tumour.

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Figure 1 Chest radiograph showing a near normal study apart from slight thickening of tissues on the left side of the thoracic inlet (arrowed).

Figure 2 MRI scan (SE-T1 coronal study) showing (A) extensive superior mediastinal and (B) left hilar lymphadenopathy with left main bronchus compression.

The patient underwent a course of systemic chemotherapy with bleomycin, vincristine, methotrexate, prednisolone, adriamycin, and cyclophosphamide (BOMPAC regimen). Folinic acid was also given in order to minimise the toxic side effects of methotrexate. Response of the tumour after six cycles of chemotherapy was excellent with resolution of the apical mass and no evidence of hilar or mediastinal nodes on CT scanning. The Horner's syndrome was still present, but the brachial neuralgia and left arm pain had improved significantly. The ejection systolic murmur heard on presentation had disappeared.

Discussion

The Pancoast syndrome is rare and is usually caused by a bronchogenic carcinoma which carries a poor prognosis. It should be remembered that, although this is by far the most common pathological cause, there have been reports of other causes.

Brachial neuritis has been described in Hodgkin's disease as a paraneoplastic syndrome,¹⁰ but it appears that malignant lymphoma as a direct cause of the Pancoast syndrome is extremely uncommon. It should, however, be considered as a cause of the syndrome as it is a potentially curable tumour.

Our case illustrates the advantages of non-invasive imaging (MRI) in determining the site of a lesion causing the Pancoast syndrome, and also the extent of spread of the disease to neighbouring structures.

The resolution of the aortic systolic murmur in this patient suggests that it was due to a local effect of the tumour on the left ventricular outflow tract, although this was not confirmed on echocardiography or on the MRI scan of the upper thorax.

Wherever possible a firm histological diagnosis should be obtained in patients presenting with the Pancoast syndrome as this significantly alters the treatment options and prognosis.

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