Pulmonary involvement in angio-immunoblastic lymphadenopathy

DAVID PRICE*
M.B., M.R.C.P.

RICHARD G. DENT†
M.B., M.R.C.P.

Peterborough District Hospital, Peterborough

Summary

Two patients are described who complained of dyspnoea and systemic symptoms. They were found to have angio-immunoblastic lymphadenopathy with extensive pulmonary involvement. In both cases, high dose corticosteroid therapy led to relief of symptoms and clearing of the diffuse radiographic shadowing seen in both lung fields.

KEY WORDS: angio-immunoblastic lymphadenopathy, pulmonary infiltration.

Introduction

Angio-immunoblastic lymphadenopathy (AIL) is a rare condition with characteristic histological features which predominantly affects the reticulo endothelial system. We report two cases with the unusual complication of pulmonary infiltration.

Case reports

Case 1

A 71-year-old man gave a 2-month history of dyspnoea, anorexia, weight loss, night sweats and fever. Examination revealed cachexia, a right supraclavicular fossa node and hepatosplenomegaly. His chest X-ray showed diffuse interstitial pulmonary infiltrates in the mid and lower zones, and bilateral hilar lymphadenopathy (Fig. 1a). Pulmonary function studies demonstrated a mixed restrictive obstructive deficit with a forced expiratory volume in 1 sec. (FEV_1) of 1.1 litres and forced vital capacity (FVC) of 1.85 litres. The supraclavicular fossa node was biopsied and histology showed the typical features of angio-immunoblastic lymphadenopathy. He was commenced on prednisolone, 60 mg daily, which produced a rapid symptomatic improvement. One month later he was very well, and his FEV_1 had improved to 2.0 litres and his FVC to 2.7 litres. His chest X-ray showed almost complete clearance of the

pulmonary infiltrates and disappearance of the hilar lymphadenopathy (Fig. 1b).

FIG. 1a. The admission chest radiograph demonstrating diffuse lower and mid zone shadowing and bilateral hilar lymphadenopathy.

Case 2

A 49-year-old Pakistani man was admitted to hospital with a 1-week history of productive cough, dyspnoea, weight loss and fever. He gave a past history of pulmonary tuberculosis. Examination revealed widespread lymphadenopathy, hepatosplenomegaly and bilateral basal crepitations. His chest X-ray showed diffuse bilateral basal shadowing and bilateral hilar lymphadenopathy. He was treated initially with triple anti-tuberculous therapy with no response. He developed a pronounced eosinophilia (2.1×10^9 /litre) and a variety of urticarial rashes.

Present address: *The General Infirmary, Leeds. †Papworth Hospital, Cambridgeshire.

FIG. 1b. The radiographic appearances 1 month later with complete clearing of the pulmonary shadowing and the lymphadenopathy.

Supra-clavicular node biopsy was performed: the microscopic features were those of angio-immunoblastic lymphadenopathy. He was commenced on prednisolone, 60 mg daily. There was initially a good symptomatic improvement and considerable clearing of the shadowing on his chest X-ray. However, 6 weeks after the commencement of steroids, his general condition rapidly deteriorated and he died. Permission for post mortem was refused.

Discussion

Angio-immunoblastic lymphadenopathy was first described in 1974 by Frizzera, Moran and Rappaport. It is most commonly found in males in their 5th and 6th decades. The clinical and pathological features have been reviewed in detail and include widespread lymphadenopathy, fever, chills, sweats, general malaise, pruritus and weight loss (Pruzanski, 1980). Eosinophilia, Coombs positive haemolytic anaemia and polyclonal hypergammaglobulinaemia are often present. Histology of involved lymph nodes characteristically shows proliferation of small blood vessels, immunoblasts and plasma cells.

Pleuropulmonary involvement remains a rare complication, there being only 15 cases reported previously (Pruzanski, 1980; Siegler and Winner, 1980). The main symptoms of pulmonary involve-

ment are dyspnoea and cough. Physical signs are often absent but fine basal inspiratory crackles have been reported on 2 occasions, and finger clubbing once. Radiographically, the usual appearance is a coarse reticulo-nodular shadowing commonly bilateral and predominantly in the mid and lower zones; pleural effusions are common as is hilar lymphadenopathy.

Lung histology has been described in 5 patients: there is a varying degree of alveolar fibrosis as well as a non-specific interstitial infiltrate with the presence of immunoblasts and plasma cells.

In a large series reported by Cullen *et al.* (1979), corticosteroids produced complete remission of the disease in 40% of 200 cases of AIL: combination chemotherapy with a variety of cytotoxics was more successful. There is some evidence that the response is better in those cases in which an allergic drug reaction has been implicated in the initiation of the disease (Newcom and Kadin, 1979). In the nine reported cases of AIL involving the lung in which steroid treatment has been given, one did not respond, 2 showed radiographic improvement but deteriorated clinically and died, and 6 responded well clinically and radiographically. There are no reports of the use of cytotoxic drugs in patients with pleuropulmonary involvement.

The long-term prognosis of patients with AIL is unpredictable and usually complicated by exacerbations and infections. Death may be the result of malignant transformation to an immunoblastic lymphoma or sarcoma, or the result of infection, renal failure, cardiovascular insufficiency, hepatic failure or acute pancreatitis.

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