Choroidal involvement in lymphomatoid granulomatosis

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Abstract

A 13-year-old boy with evidence of pulmonary lymphomatoid granulomatosis developed monocular diplopia. Fluorescein angiography revealed bilateral choroidal involvement. Following treatment with vincristine, cyclophosphamide, and prednisone his diplopia resolved and the angiographic appearances returned to normal.

Figure 1 Chest radiograph showing bilateral diffuse infiltrates.

Figure 2 Lung biopsy showing characteristic appearances of lymphomatoid granulomatosis. (H-E, $\times 135$.)

Lymphomatoid granulomatosis was first described by Leibow *et al* in 1972.¹ It is a form of angitis with granulomatosis which predominantly affects the lungs, skin, nervous system, and kidneys. The condition is a multisystem disease with a mortality rate of up to 63%, death being due either to respiratory failure or to the development of malignant lymphoma. Characteristically the patient presents with widespread pulmonary infiltrates, dyspnoea, and respiratory failure. The central nervous system is involved in 40% of patients. Little, however, has been described of ophthalmic manifestations of the disease. It appears to be uncommon in Europe and unusual in children. The initial presentation of this boy has been previously reported.²

Case report

At the age of 10, in 1981, this boy presented with a productive cough, fever, night sweats, and lethargy. Despite antibiotics he developed respiratory failure and cyanosis, dyspnoea, intercostal and subcostal recession, and crepitations in both lung fields. The liver and spleen were both palpable. A radiograph of his chest showed bilateral dense shadows (Fig 1). Other investigations gave normal results. Respiratory failure progressed, and he required mechanical ventilation. In view of this an open lung biopsy was performed. Macroscopically the lung was nodular, and histologically there were dense interstitial infiltrates of lymphocytes, polymorphs, and histiocytes (Fig 2).

These cells invaded the walls of many arteries with consequent obliteration, and there were many areas of necrosis and peripheral fibrosis. These are the features of lymphomatoid granulomatosis. He was treated with 60 mg of prednisone daily and made a very rapid recovery. However, withdrawal of steroids resulted in the return of his respiratory symptoms, and he was therefore kept on a small maintenance dose.

In May 1982 he developed bilateral parotid swellings and right facial palsy. A biopsy showed infiltration with lymphocytes and plasma cells, with a disordered arrangement of salivary gland ducts. Once again treatment with high dose prednisone resulted in a rapid resolution of these masses within 10 days. Concurrent skin infiltrates also disappeared. In November 1983 while on 10 mg of prednisone daily he developed monocular diplopia, and acute dizziness. A chest x ray at this time was normal as were the findings on general examination, apart from a raised blood pressure of 180/110 mm Hg. Investigation of his hypertension revealed that there was scarring of both kidneys on a dimercaptosuccinic acid scan and raised serum renin and aldosterone concentrations, although an intravenous pyelo-

Figure 3 Fluorescein angiogram showing hyperfluorescent choroidal lesion in the lower temporal quadrant of the left eye.¹

gram and ultrasound picture were normal. This was thought to be consistent with previous involvement of his kidney with lymphomatoid granulomatosis.

Ophthalmic examination showed the cornea to be clear, and there was no evidence of lens opacities. Funduscopy showed an area of retinal oedema just below the macula. Fluorescein angiography showed a hyperfluorescent choroidal lesion just below and temporal to the macula of the left eye (Fig 3). The findings were consistent with a choroidal focus of lymphomatoid granulomatosis.

In view of the widespread nature of his disease he was started on vincristine, cyclophosphamide, and prednisone. Within two weeks his diplopia resolved, and a fluorescein angiogram performed one month later was entirely normal. Six years later he has had no recurrence of the ocular symptoms.

Discussion

Lymphomatoid granulomatosis is a multisystem disorder characterised pathologically by a peri-

vascular infiltrate of lymphocytes. Many organs may be affected but predominantly the lungs, central nervous system, and kidneys. This boy presented with monocular diplopia, and fluorescein angiography showed multiple choroidal involvement. Diplopia has been previously recorded in this condition secondary to lesions in the central nervous system and cranial nerves. Choroidoretinal involvement has been described in a 29-year-old male³ and in two men aged 42 and 55 years.⁴ Two of the patients previously reported had no ocular symptoms seven and 10 years after the onset, and the other had died with systemic involvement by the disease. Fauci *et al*⁵ suggested that vincristine, cyclophosphamide, and prednisone were beneficial in this disease and caused more improvement than steroids alone. The boy described here responded dramatically to treatment with this combination, and his infiltrates disappeared.

Fluorescein angiography displayed the infiltrates in his choroid at a stage when routine funduscopy showed only retinal oedema.

This case again demonstrates that the choroid is yet another organ which can be involved with lymphomatoid granulomatosis. Fluorescein angiography aids its diagnosis, and treatment with vincristine, cyclophosphamide, and prednisone produced a dramatic and long lasting response in this child.

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