Lymphomatoid granulomatosis presenting with ornithosis pneumonia

Lymphomatoid granulomatosis is a proliferative disorder of unknown aetiology that primarily affects the lungs but may also affect the skin, nervous system, and kidneys. It was first described in 1972 by Liebow et al. and is characterised histologically by an angiocentric, angiodestructive infiltrate of atypical lymphocytes and histiocytes. We describe the sixth case of lymphomatoid granulomatosis to be reported in the United Kingdom; ornithosis pneumonia was a concurrent finding.

Case report

A 34 year old woman was admitted in 1979 with a three month history of cough and weight loss. One week before admission she had developed dyspnoea, fever, and pleuritic chest pain. On examination she was feverish (39°C) and had late inspiratory crackles in the right lower zone of the chest and weakness of the right quadriceps. A chest x ray film showed widespread bilateral fluffy pulmonary opacities (figure).

Atypical pneumonia was presumptively diagnosed and treatment with erythromycin started. Fibreoptic bronchoscopy the next day showed normal bronchi, but transbronchial biopsy specimens showed swelling of the epithelial cells of the alveolar wall, suggesting ornithosis pneumonia. Prolonged Giemsa staining showed intracytoplasmic inclusions (Levinthal-Coles-Lillie bodies) typical of ornithosis. Titres of psittacosis and lymphogranuloma venereum antibodies in the serum were greater than 1/100 and subsequently fell to 1/40. Her only contact with birds had been with a budgerigar; after its death its tissue was examined but did not show any evidence of infection.

Antibiotic treatment was changed to doxycycline, and after three weeks her temperature settled but she remained dyspnoeic and developed finger clubbing. Pulmonary function tests showed a restrictive ventilatory defect. Transbronchial biopsy specimens taken at six weeks showed considerable interstitial fibrosis. Steroid treatment was started with 30 mg prednisolone daily.

Antibiotics were stopped after two months; at three months she was clinically fully recovered and a chest x ray film was normal. Steroid treatment was gradually reduced and stopped after four months. She remained well until six months later, when a single pulmonary nodule 2 cm diameter developed in the right lung and an abdominal mass was found in the right upper quadrant. Laparotomy showed a lesion affecting the small bowel and mesenteric lymph nodes, which was resected. Macroscopically the appearances were those of a lymphoma, and lymphomatoid granulomatosis was confirmed by histological examination.

Prednisolone was started again at 60 mg daily; the opacity in the chest x ray film cleared in four months, and the prednisolone dosage was slowly reduced. She remained well three years later and was taking 5 mg prednisolone.

Comment

Although lymphomatoid granulomatosis is a recognised cause of pyrexia of unknown origin, the diagnosis of ornithosis infection was confirmed by histological and serological examination. The initial radiological appearance (figure) was compatible with parenchymal pulmonary lymphomatoid granulomatosis but would be more consistent with severe ornithosis pneumonia. This can also progress to pulmonary fibrosis, which sometimes responds to steroids. The subsequent nodular opacity, however, was typical of pulmonary lymphomatoid granulomatosis. Isolated peripheral neuropathy may rarely complicate ornithosis but is common in lymphomatoid granulomatosis. It is therefore difficult to know whether ornithosis pneumonia preceded the development of lymphomatoid granulomatosis or was a coexistent opportunistic infection.

Cyclophosphamide is often administered with corticosteroids in the management of lymphomatoid granulomatosis but was not used in this case because of the initial good response to steroids and the risk of precipitating an overwhelming infection. It has been suggested that lymphomatoid granulomatosis confers a worse prognosis when it affects the gastrointestinal tract. The average two year survival of patients with the condition is less than 40%, and it is therefore interesting that our patient responded to steroids alone.

Lymphomatoid granulomatosis is a disease in which reactive and neoplastic changes merge. The possible coexistence of an intracellular infection should therefore not be ignored. Although Chlamydia psittaci was probably an opportunistic coexistent infective agent, an aetiological role cannot be completely excluded. Epstein-Barr virus has already been implicated in this respect. We therefore suggest that all patients with lymphomatoid granulomatosis should be comprehensively screened for infective agents.

We thank Professor B Heard, of the Brompton Hospital, London, Professor D Turner, of the University Hospital, Nottingham (formerly of Musgrove Park Hospital, Taunton), and Dr N Rankin, of Yeovil District Hospital, for their expert opinions on the histopathological findings in this case.


(Accepted 9 December 1983)

Yeovil District Hospital, Yeovil, Somerset BA21 4AT

R M GRAHAM, MB, MRCP, registrar in medicine
I W FAWCETT, MA, MRCP, consultant physician

Correspondence to: Dr R M Graham, registrar in dermatology, Royal Berkshire Hospital, Reading, Berks RG1 5AN.

SPIRIT OF CASTOREUM—"Take of fresh Castoreum four ounces, Lavender flower an ounce, the tops of Sage and Rosemary, of each half an ounce, Cinnamon six drams, Mace, Cloves, of each two drachms, spirits of Wine rectified, six pounds, digest them in a phial filled only to the third part, close stopped with cork and bladder in warm ashes for two days, then distilled in Balneo Mariae, and the distilled water kept close stopped."

By reason of its heat it is no ways fit to be taken alone, but mixed with other convenient medicines appropriated to the diseases you would give it for, it resists poison, and helps such as are bitten by venomous beasts: it causes speedy delivery to women in travail, and casteth out the Placenta: it helps the fits of the mother, larynges and convulsions, being mixed with white wine, and dropped into the ears, it helps deafness; if stopping be the cause of it, the dose to be given inwardly is between one dram, and half a dram, according to the strength and age of the patient. (Nicholas Culpeper (1616-54) The Complete Herbal, 1650.)