Case report

An unusual case of systemic lupus erythematosus with isolated hypoglossal nerve palsy, fulminant acute pneumonitis, and pulmonary amyloidosis

C N CHAN, E LI, F M LAI, AND J A PANG

From the Department of Medicine and Department of Morbid Anatomy, The Chinese University of Hong Kong, Prince of Wales Hospital, Shatin, NT, Hong Kong

SUMMARY A 53 year old Chinese man with systemic lupus erythematosus (SLE) had an isolated 12th cranial nerve palsy, and acute pneumonitis. He died of respiratory failure despite intensive treatment. A limited necropsy was performed, and amyloid deposits were identified in both lung and kidney tissue. This case is highly unusual because (a) to our knowledge an isolated hypoglossal nerve palsy associated with active SLE has never been reported; (b) only one of nine reported cases of amyloidosis in patients with SLE had amyloid deposits in the lung.

Both acute pneumonitis\(^1\) and mononeuropathy\(^2\) are rare manifestations of systemic lupus erythematosus (SLE), and an isolated 12th cranial nerve palsy has not been described previously. The presence of these features and the additional necropsy finding of pulmonary amyloidosis in a male patient make this case unique. There are only a few reports of amyloidosis associated with SLE.\(^3\) Despite reports of acute lupus pneumonitis responding favourably to immunosuppressive treatment,\(^4\) our patient succumbed without evidence of severe infection. We suggest that pulmonary amyloidosis may have contributed to his rapid demise.

Case report

A 53 year old Chinese man was admitted in January 1986 with a year’s history of fever, weight loss, multiple joint pains, and dyspnoea. On admission he was febrile (39°C) with a prominent malar rash, angular stomatitis, and non-deforming polyarthritis involving both hands with periungual infarcts. Inspiratory crackles were audible at both lung bases. In addition, there was an isolated right 12th cranial nerve palsy with a wasted, fibrillating tongue.

Four months later he was readmitted with a month’s history of fever, weight loss, multiple joint pains, and dyspnoea. On admission he was febrile (39°C) with a prominent malar rash, angular stomatitis, and non-deforming polyarthritis involving both hands with periungual infarcts. Inspiratory crackles were audible at both lung bases. In addition, there was an isolated right 12th cranial nerve palsy with a wasted, fibrillating tongue.

Initial investigations showed the following: haemoglobin 130 g/l, leucocyte count 6-4x10^9/l (lymphocytes 0-9x10^9/l), platelet count 186x10^9/l, erythrocyte sedimentation rate 76 mm/h (Westergren), normal renal and liver function tests. Urine analysis was normal. Globulin was raised at 68 g/l (normal 30–33) with low albumin 290 g/l (normal 360–480). Rose-Waaler test (Venereal Disease Research Laboratory) and hepatitis B surface antigen were negative. Antinuclear antibody was weakly positive with a titre of 1/40. Anti-DNA was undetectable. C3 and C4 concentrations were normal. Arterial blood gas measurements showed a PaO\(_2\) of 8 kPa and PaCO\(_2\) of 4 kPa on admission. A chest radiograph showed high diaphragms with patchy shadowing in both bases. Pulmonary function tests gave the following results: forced expiratory volume in one second (FEV\(_1\)) 1.91 litres (68% of predicted value), forced vital capacity (FVC) 2.42 litres.
litres (65% of predicted value), FEV₁/FVC ratio 78%, and transfer factor of the lung for carbon monoxide 15.4 ml/min/mmHg (normal 21.2 ml/min/mmHg), 72% of predicted value.

After admission he continued to have swinging fever and developed widespread crackles over both lung fields with increasing hypoxaemia. Repeat chest x rays showed extensive infiltrates. Screening for atypical organisms, opportunistic infections, and viruses was negative. An echocardiogram showed a mild pericardial effusion with normal ejection fraction. A transbronchial lung biopsy showed chronic inflammation within alveoli with repair and hyperplasia of pneumocytes typical of interstitial pneumonitis. Focal areas of acute fibrin and hyaline exudation with blood clots were also evident. There was no apparent vasculitis. ‘Pulse’ treatment with 1 g methylprednisolone for three days followed by high dose oral prednisone (80 mg/day) was started 12 days after admission.

Despite treatment the patient failed to respond. Repeat lung function three weeks after admission showed further deterioration: FEV₁ 1.75 litres (62% of predicted value), FVC 1.96 litres (52% of predicted value), and FEV₁/FVC ratio 89%. Five weeks after admission he required ventilatory support for respiratory failure (Pao₂ 6 kPa and Paco₂ 4 kPa on 6 litres/min oxygen inhalation). Cyclophosphamide was added, but the patient died one week later.

Permission for a full necropsy was not granted, and examination was limited to the heart, lung, spleen, kidneys, and liver.

The postmortem findings were as follows: (a) light microscopy of both lungs showed acute interstitial reactions with diffuse alveolar damage (Fig. 1). No vasculitic lesions were seen. Deposits of an amorphous eosinophilic substance were found in many vessels, which stained positively for Congo red and was green birefringent under polarised light (Fig. 2). (b) An immunofluorescent study of both kidneys confirmed the presence of segmental glomerular
deposition of IgM, IgA, C1q, C3, and C4. Focal segmental positive amyloid staining was also identified within a few glomerular capillary loops. There were no significant findings in the heart, liver, and spleen.

Discussion

A clinical diagnosis of SLE was made in this patient on his second admission with the presence of a malar rash, stomatitis, pericarditis, polyarthritis, and recurrent lymphopenia, fulfilling the revised American Rheumatism Association Criteria for the classification of SLE. He had additional features of cranial nerve palsy, digital vasculitis, and pneumonia.

This patient is highly unusual in several aspects. Firstly, although neuropsychiatric manifestations in SLE have been frequently reported, mononeuropathy is uncommon, varying from 2% to 29% of cases. In our search of the English published work we did not find any report of an isolated 12th cranial nerve palsy, and our patient may be the first case with this manifestation. Pathological findings suggest that mononeuropathy is a vasculitic phenomenon of SLE. Sadly, we were unable to seek postmortem evidence of the pathogenesis of the nerve palsy.

Secondly, amyloid was found in the lungs and kidney. Nomura et al recently described the first case of pulmonary amyloidosis associated with SLE and reviewed eight other previously reported cases, all of whom had significant renal amyloidosis. Our patient had both renal and pulmonary amyloidosis.

Thirdly, the preceding history of intermittent haemoptysis and pleuritic pain for one year suggests that pleuropulmonary complications had already been present for some time despite the absence of...
other clinical features. Although 50–70% of cases of SLE are reported to have some degree of pleuropulmonary involvement, this mode of presentation is unusual. Moreover, the patient developed acute lupus pneumonitis during his final admission, and this is also uncommon, being present in only 11–7% in the series of Matthay et al.1

Smith et al reviewed 232 necropsy files with a diagnosis of amyloidosis and reported pulmonary involvement in 30%, mainly in senile and systemic amyloidosis, and concluded that despite its presence, amyloidosis was largely inconsequential and caused no significant pulmonary impairment.10 Our patient died in respiratory failure despite being treated with steroids and cyclophosphamide, which are said to be dramatically effective in acute lupus pneumonitis.4 5 Despite the 50% mortality reported in the review of 12 such cases by Matthay et al,1 only three died of respiratory failure, while the others succumbed to causes unrelated to their pulmonary disease. It is possible that the lack of response may be partly related to pre-existing chronic lung disease. Thus amyloidosis may have contributed to his rapid demise, and the view that pulmonary amyloidosis is largely inconsequential10 may not apply to this condition when it is associated with SLE.

References