LETTERS TO THE EDITOR

Laryngeal involvement as a presenting symptom of systemic lupus erythematosus

Sir: The actual prevalence of laryngeal involvement in systemic lupus erythematosus (SLE) is unclear, but it seems to be very uncommon. Furthermore, laryngeal vasculitis in SLE is even rarer and has not yet been reported. In fact, in the lupus erythematosus textbook of Wallace and Dubois no mention is made of the actual prevalence and type of damage of the larynx. We have recently seen a young man with SLE who developed hoarseness as the first symptom.

A 33 year old white man was admitted in December 1988 with a five week history of hoarseness. Three weeks later he noticed anorexia, fatigue, and purpuric, palpable purpura, and paraesthesia in the legs. Before admission the patient had only received non-steroidal anti-inflammatory drugs, without improvement. His medical history was uneventful.

Initial examination showed a normal blood pressure, a temperature of 37°C, and a pulse of 90 beats a minute. Polyarthritus, oral ulcers, and palpable purpura in the legs were noticed.

A direct laryngoscopy showed complete paralysis of the right vocal cord without masses and with no evidence of external compression. Laboratory investigation showed a haemoglobin of 132 g/l and a white blood count cell of 9-7 x 10^3/l with 1-261 x 10^9/l lymphocytes. The erythrocyte sedimentation rate was 49 mm/hour (Wintrobe). The blood chemistry was normal, with a urea of 6.5 mmol/l and the serum creatinine was 159 µmol/l with a normal glomerular filtration rate. A chest x ray was normal. Four weeks after admission the creatinine rose to 451 µmol/l and the glomerular filtration rate dropped to 42 ml/min. Urine analysis showed innumerable red blood cells and 14 white blood cells per high power field, and granular and hyaline casts. Albuminuria of 2.49 g in 24 hours was noted. Immunological studies showed antinuclear antibodies and anti-double-stranded DNA antibody.

A kidney biopsy was done and showed a diffuse proliferative glomerulonephritis with an activity index of 20 and chronicity index of 6. A skin biopsy showed a leukocytoclastic vasculitis, and a nerve conduction study was compatible with a peripheral neuropathic pattern.

The patient was treated initially with three 'bolus' of intravenous methylprednisolone (1 g/day). One month later, because of further evidence of kidney damage, treatment was started with intravenous cyclophosphamide, while treatment was continued with 60 mg prednisone in one dose. Six months later the serum creatinine was 186 µmol/l. The hoarseness and the paraesthesia improved rapidly after the initial methylprednisolone bolus.

Systemic lupus erythematosus was diagnosed in our patient according to the 1982 revised criteria—that is, he had non-erosive arthritis, oral ulcers, lymphopenia, kidney damage, and positive antinuclear antibodies. 1-3 The prevalence of masses or extrinsic compression was found at laryngoscopy and the chest x ray was normal, excluding pulmonary hypertension, we concluded that vasculitis was the underlying mechanism of this patient's hoarseness. We were unable to exclude the possibility that neuropa thy involvement of the recurrent laryngeal nerve was the underlying mechanism of damage. We feel, however, that the concomitant appearance of vasculitis in the skin shown by skin biopsy and the prompt resolution of dysphonia following the administration of the methylprednisolone suggests that vasculitis was the mechanism of damage.

Laryngeal nerve palsy as a manifestation of SLE is an uncommon phenomenon and to the best of our knowledge it has not been reported as the initial manifestation. In the report of Asherson et al of pulmonary hypertension in SLE one patient had vocal cord palsy which might have been vasculitic in origin. The same authors in 1985 described two patients with vocal cord paralysis in SLE secondary to pulmonary hypertension, excluding vasculitis as the cause of the dyspnoea. Other authors have reported recurrent laryngeal nerve palsy secondary to direct compression of the nerve by a dilated pulmonary artery. 4 We feel that the hoarseness in our patient was not secondary to pulmonary hypertension because of the normal chest x ray and because of the rapid disappearance of the hoarseness following the methylprednisolone.

Undoubtedly, SLE is a disease with many protein and occasionally unpredictable symptoms and signs, which may form part of the disease spectrum. We suggest that persistent hoarseness should be added to these.

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Childhood adrenal insufficiency, chorea, and antiphospholipid antibodies

Sir: Carette and Jobin in a recent issue of the Annals reported a patient with adrenal insufficiency, deep vein thrombosis, and antiphospholipid antibodies.

The child was an 11 year old girl who presented with cutaneous and mucosal purpura and with oral and periorbital edema. The patient had a history of recurrent deep vein thrombosis, pulmonary embolism, and recurrent pregnancy loss. Laboratory tests revealed a normal serum cortisol level and an adequate response to cosyntropin. Antiphospholipid antibodies were present in the patient and her mother.

Secondary adrenal insufficiency is considered an emerging clinical syndrome. Systemic lupus erythematosus was diagnosed in seven of the 20 children with antiphospholipid antibodies. These children were treated with low dose oral prednisone and showed rapid resolution of their clinical manifestations. We were able to discontinue all immunosuppressive therapy in all these children. The child presented had no history of recurrent deep vein thrombosis, pulmonary embolism, or thrombocytopenia. We were unable to establish a definite diagnosis of antiphospholipid syndrome.

A 15 year old white girl was admitted to the Children's Hospital of Philadelphia in March 1989 with a recent diagnosis of Addison's disease. She subsequently developed chorea without other clinical features of systemic lupus erythematosus. A high positive antinuclear antibody titre (1/1280) prompted consultation with the rheumatology service. Further investigations showed the absence of antibodies to dsDNA, extractable nuclear antigen, and adrenal tissue, a prolonged partial thromboplastin time, and highly increased levels of IgM anticardiolipin antibody (enzyme linked immunosorbent assay (ELISA)), the IgG anticardiolipin antibody titre was slightly raised. She was treated with haloperidol and corticosteroids (1 mg/kg daily). She improved neurologically and tolerated a slow tapering of corticosteroid treatment in the following weeks. This patient, except for her age, is similar to those already reported. 1, 2

Primary adrenal failure is very rarely seen in systemic lupus erythematosus, 3 and chorea is a well recognised, but also a rare manifestation. We feel that this young girl represents a paediatric example of the 'primary antiphospholipid syndrome'. 4 In a recent series of 12 patients with chorea Asherson et al noted a striking association with antiphospholipid antibodies. 5 Six (50%) of these patients were defined as having 'lupus like' syndromes owing to a lack of defined criteria for systemic lupus erythematosus. 6 Our patient would seem to belong to this subtype.

The main cause of adrenal insufficiency is autoimmune destruction of the adrenal glands. Two thirds of patients with Addison's disease have antirenal autoantibodies in their sera. 7 The cause of the disease in the remaining one third is less well defined but includes chronic infections and granuloma. It is of great interest that possibly a new 'subset' may be emerging within the group of patients with primary antiphospholipid syndrome, and it is clearly of interest to search for these antibodies in patients with primary adrenal failure, even if their pathophysiological significance is still unclear.

Patients with and without systemic lupus erythematosus with positive antiphospholipid antibodies often have thrombotic complications, but a pathogenetic role for those antibodies has not been established as yet. In some patients with adrenal failure a 'true' cause has been suggested. 1, 2, 7 An antibody mediated block in steroid synthesis should possibly also be considered. In vitro studies might be helpful in answering this question.

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