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.1 Furthermore, laryngeal vasculitis in SLE is even rarer and has been scarcely reported.2 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, weight loss, oral ulcers, arthritis, 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. An indirect laryngoscopy showed complete paralysis of the right vocal cord without masses and with no evidence of external compression.

Laboratory investigations showed a haemoglobin of 132 g/l and a white blood cell count of 9.7 × 10^9/l with 1:261 × 10^9/l lymphocytes. The erythrocyte sedimentation rate was 49 mm/hour (Wintrobe). The blood chemistry was normal, except for a creatinine level of 159 μmol/l and the serum creatinine was 159 μmol/l and a normal globular 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 evident. Immunological studies showed antinuclear antibodies and double-stranded DNA antibody.

A kidney biopsy was done and showed a diffuse proliferative glomerulonephritis with an activity index of 20 and a chronicity index of 6. A skin biopsy showed a leucocytoclastic 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.2 As an option, the presence 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 neuropathy without other clinical neurological signs or symptoms in 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 dysphonia. Other authors have reported recurrent laryngeal nerve palsy secondary to direct compression of the nerve by a dilated pulmonary artery.3 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: Carterre and Jobin in a recent issue of the Annals reported a patient with adrenal insufficiency, deep vein thrombosis, and antiphospholipid syndrome. In this patient systematic lupus erythematosus was diagnosed, and the patient had been treated with rituximab. The authors concluded that the antiphospholipid antibodies might have induced an autoimmune mechanism, which might have been an initiating factor in the development of the systemic lupus erythematosus.

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 manifestations of systemic lupus erythematous. A high positive anti-nuclear antibody titre (1/1280) prompted consultation with the rheumatology service. Further investigations showed the absence of antibodies to dsDNA, and anti-CL, while the patient had antibodies to Ro/SS A and AS/SS B. A positive anti-cardiolipin antibody (an IgG) was found in the presence of an anti-CL antibody. The patient also had an elevated erythrocyte sedimentation rate of 90 mm/hr, a high C3 of 33 mg/dl, and a high C4 of 90 mg/dl. She also had a low platelet count of 20000. Her antinuclear antibodies were positive at a titre of 1/1280. Tests for dsDNA, and anti-CL, while the patient had antibodies to Ro/SS A and AS/SS B. A positive anti-cardiolipin antibody (an IgG) was found in the presence of an anti-CL antibody. The patient also had an elevated erythrocyte sedimentation rate of 90 mm/hr, a high C3 of 33 mg/dl, and a high C4 of 90 mg/dl. She also had a low platelet count of 20000. Her antinuclear antibodies were positive at a titre of 1/1280. Tests for anti-dsDNA, anti-CL, and anti-SS B were positive at titres of 1/1280, 1/640, and 1/640, respectively.

The main cause of adrenal insufficiency is autoimmune destruction of the adrenal glands. Two thirds of patients with Addison's disease have adrenocortical autoantibodies in their sera.4 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 pathophysiologic significance is still unclear.

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

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