LETTERS TO THE EDITOR

Progressive systemic sclerosis with CNS vasculitis and cyclosporin A therapy

Progressive systemic sclerosis is a multi-system disorder characterised by inflammatory, vascular and fibrotic changes of skin and a variety of internal organs. Most reports describing neurological complications of PSS have focused on damage to the peripheral nervous system. CNS dysfunctions are rare and are thought to result from uraemia, hypoxaemia, and severe hypertension. This is the first trial of cyclosporin A (CyA) in a patient with CNS vasculitis secondary to PSS.

A 67 year old woman was admitted with difficulty in walking and with speech. She had been in good health until December 1985, when she noted that her hands turned white in a cold environment. Subsequently, she developed a nonproductive cough. In February 1986 she experienced occasional dizziness. In July 1990 she began to walk with a waddle.

On admission, her blood pressure, pulse, body temperature, and respiration were normal. The skin of her face was so taut that she was unable to open her mouth fully; her lips were thin and shortened. She had telangiectasia on the chest. Auscultation of the chest disclosed fine crackles on the lower fields of both lungs. The skin of her fingers and hands was firm and thickened and tightly bound to underlying subcutaneous tissue. The skin changes spread from her hands to the chest. Calcinosis cutis was not found. Raynaud’s phenomenon could be induced by cold stimuli.

She had dysarthria; left hemiparesis with slight rigidity and spasticity; and mild incoordination. Deep tendon reflexes were normal and pathological reflexes were absent.

The ESR, blood count, and biochemical tests, were normal. The C-reactive protein was negative; rheumatoid factor positive; antinuclear antibody present in a 1:320 dilution; anti-ScI-70 antibody (antiscieroderma antibody) was positive in a 1:16 dilution.

Electrocardiogram showed first-degree atrioventricular block with a prolonged PR interval of 0-24 second. Pulmonary function tests revealed a slightly restrictive ventilatory defect; the ratio of FEV1/FVC was 0.94 and the ratio of FVC observed/FVC predicted was 0.73. Cold water (10°C) immersion test on the skin of the hand showed a delay in return to the initial temperature, which was compatible with Raynaud’s phenomenon. CSF was normal. Radiograph of the chest showed fine fibrotic strands in the lower portions of both lungs. Barium-meal revealed a dilated oesophagus with decreased peristalsis. MRI scan of the head disclosed ischaemic lesions in the right occipital region, the right middle cerebellar peduncle, and the basilar part of the pons.

Digital subtraction angiography (DSA) showed localised severe stenosis and post-stenotic dilation, a "string of beads" appearance, from the distal end of the right vertebral to the proximal portion of the basilar arteries (fig). There was no definite stenosis in other arteries. Skin biopsy specimens showed proliferation of thickened collagen fibres in the dermis.

Based on these findings, we diagnosed PSS, following the scleroderma criteria of the American Rheumatism Association, and attributed the abnormal angiogram findings to vasculitis. Treatment was started with CyA 8 mg/kg body weight on admission. Within weeks later, urinalysis disclosed an increase in urinary β2-microglobulin, a sign of renal damage caused by CyA. Immediately the dose was reduced to 6 mg/kg, and during the next month it was reduced by 1 mg/kg every week, to 3 mg/kg.

Though the abnormal findings of serological tests were not improved, during this period the β2-microglobulin returned to normal. The patient's nonproductive cough subsided and Raynaud's phenomenon did not occur. The skin involvement gradually diminished, and the abnormal ECG findings and pulmonary function tests improved: PR interval, 0-20 second; FVC observed/FVC predicted ratio, 0-8. The neurological symptoms that resulted from multiple CNS infarction remained stable. Cold water immersion test showed a normal recovery to the initial temperature.

One year after the beginning of CyA treatment, the patient takes 150 mg/day (3 mg/kg/day) of CyA, her hands and fingers are still slightly thickened. Two months later, urine analysis disclosed an increase in urinary β2-microglobulin, a sign of renal damage caused by CyA. Immediately the dose was reduced to 6 mg/kg, and during the next month it was reduced by 1 mg/kg every week, to 3 mg/kg.

In this study, we verified the diagnostic sensitivity of single fibre EMG of the orbicularis oculi muscle in patients with ocular myasthenia gravis. We found that increased jitter and impulse blocking can also be found in nerve and muscle diseases, and abnormal findings have been reported in some patients with chronic progressive external ophthalmoplegia.

When myasthenia gravis is restricted to ocular muscles it can present a difficult diagnosis. Firstly, the response to Tensilon iv can be negative or ambiguous; secondly, repetitive nerve stimulation and titration of serum anti-acetylcholine receptor antibodies (anti-AChR abs) often give negative results. Chronic progressive external ophthalmoplegia (CPEO) must be distinguished. Single fibre electromyography (SFEMG) has proved to be a useful diagnostic test in patients with myasthenia gravis. However, increased jitter and impulse blocking can also be found in nerve and muscle diseases, and abnormal findings have been reported in some patients with chronic progressive external ophthalmoplegia.

Ocular myasthenia: diagnostic value of single fibre EMG in the orbicularis oculi muscle

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