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Cerebellar atrophy in systemic sclerosis

Luís Pinheiro MD¹ João Freitas MD²
Margarida Lucas MD¹ Rui M M Victorino MD PhD¹

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Cerebellar atrophy has been recognized as an occasional feature of systemic lupus erythematosus (SLE)^{1,2} but not systemic sclerosis.

CASE HISTORY

The patient was a woman whose illness had begun at the age of 40 with Raynaud's phenomenon, progressing to distal necrosis and ulceration of fingertips, with sclerosis of the hands, feet and face. The results of investigation were consistent with systemic sclerosis (SS). Subsequently she reported heartburn and regurgitation, and oesophageal manometry revealed a defect compatible with SS. Findings on oesophagogastroscopy and echocardiography were normal; respiratory function tests showed slight ventilatory dysfunction. At age 46 she sought advice after six months of progressive unsteadiness and ataxia. On examination her gait was broad-based and she had cephalic and intention tremors with bilateral dysmetria. Motor and sensory nerve function was normal. Tests for antinuclear, anticentromere and anti-SSA antibodies were positive; those for anti-SSB, anti-dsDNA, anti-Jo1, anti-Scl70 and anti-RNP were negative. She did not report dry eyes or dry mouth and Schirmer's test was negative. The echocardiogram and the respiratory function tests had not changed. Cranial CT (Figure 1) revealed severe diffuse symmetrical cerebellar atrophy without evidence of demyelination or infarction; no lacunae (basal or cortical) were present and the

Figure 1 Cranial CT showing cerebellar atrophy

supratentorial subarachnoid space was normal; there were no features suggestive of vascular disease and the carotid and vertebral arteries showed no calcifications. The patient did not use alcohol and did not recall any exposure to toxic substances; investigations for possible underlying occult neoplasia were negative. After methylprednisolone pulse therapy (1 g per day for three days) the cerebellar signs improved and she became able to walk without assistance, though the ataxia and unsteadiness did not resolve completely. A year later, pulmonary hypertension was diagnosed (pulmonary artery systolic pressure 58 mm Hg) with depression of right ventricular function, dilatation of the right atrium and tricuspid insufficiency. Helical thoracic CT showed no evidence of vascular thrombi or pulmonary fibrosis and the ventilation–perfusion scan did not support the diagnosis of pulmonary embolism. The pulmonary hypertension was interpreted as secondary to the pulmonary microvascular disease of systemic sclerosis. Intravenous Iloprost (a prostacyclin analogue) decreased the pulmonary systolic pressure to 41 mm Hg and she improved clinically. She was then switched to inhaled Iloprost and the improvement continued after discharge. The cerebellar ataxia was no worse than it had been a year before. A month after discharge, she was readmitted severely hypoxic with pneumonia and had an irreversible cardiorespiratory arrest.

Departments of ¹Internal Medicine II and ²Dermatology, Hospital Santa Maria, Lisbon, Portugal

Correspondence to: Professor Rui M M Victorino, Serviço de Medicina 2, Hospital de Santa Maria, Av Prof Egas Moniz, 1649-028 Lisboa, Portugal

E-mail: rvict@fm.ul.pt

COMMENT

This patient had typical systemic sclerosis with skin and oesophageal involvement and progressive pulmonary hypertension. The unusual clinical feature was the ataxia due to cerebellar atrophy. The diagnosis of systemic sclerosis was supported by the finding of anticentromere antibodies (associated with the development of non-fibrotic pulmonary hypertension in that condition³) and antinuclear antibodies (present in more than 70% of patients^{3,4}); the anti-SSA antibodies were also consistent with the diagnosis.^{3,5} Progressive subacute cerebellar degeneration can be part of a paraneoplastic syndrome, but there was no evidence of neoplasia on investigation or subsequent follow-up.

In SLE, central nervous system disease is frequent whereas in systemic sclerosis it is rare⁶ and apparently unrelated to systemic vascular damage.⁷ We have found no previous reports of cerebellar atrophy associated with systemic sclerosis. The features resembled those in occasional cases of SLE, as did the response to treatment. Al Arfaj and Naddaf¹ saw one patient with cerebellar ataxia in a series of 150 with SLE and this patient responded to high-dose steroids and azathioprine; Singh *et al.*² detected cerebellar signs in 3 of 350 SLE patients, and these patients too improved on steroid therapy. The mechanism in some patients may be immunological. Tomita⁸ identified in an animal model of SLE a specific cytokine profile with enhanced gene expression of interleukin (IL)-6, interferon-gamma, IL-1 β and IL-10 associated with the development of cerebellar signs. We likewise speculate that the cerebellar involvement in our patient with systemic sclerosis was a manifestation of autoimmunity.

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Cryptogenic multifocal ulcerous stenosing enteritis

Hal Spencer MRCP¹ Panagiota Kitsanta PhD MD²
Stuart Riley ChB FRCP¹

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A patient with anaemia was initially thought to have gastrointestinal cancer and subsequently Crohn's disease, but pathological examination revealed a much rarer condition.

CASE HISTORY

A woman of 66 was referred for investigation of colicky lower abdominal pain, iron deficiency anaemia and weight loss of 7 kg in the previous nine months. For much longer she had suffered from constipation. A Paget's carcinoma of the breast had been treated by mastectomy 4 years earlier but there was no clinical evidence of recurrence. She had mild chronic obstructive pulmonary disease.

On examination she was pale and unwell. Her abdomen was distended but there was no peritonism. Investigations confirmed iron deficiency anaemia with haemoglobin 9.8 g/dL and ferritin 2 μ g/L. Her platelet count was also slightly raised at $395 \times 10^9/L$ and her erythrocyte sedimentation rate (ESR) was 32 mm/h. Faecal occult bloods were strongly positive. At gastroscopy she was found to have gastroduodenitis but duodenal biopsies were normal. A barium enema revealed no lesions of note. She was initially reassured but her haemoglobin then fell to 9.0 g/dL. Colonoscopy was therefore undertaken but revealed only haemorrhoids and diverticulosis. Colonic and terminal ileal biopsies were normal. Despite oral iron therapy, the haemoglobin fell further to 6.7 g/dL (MCV 56 fL) and her ESR remained raised at 54 mm/h. She was admitted for transfusion and further investigation. Ultrasound revealed 'to-and-fro peristalsis in fluid-filled bowel consistent with intestinal obstruction'. CT confirmed the presence of small-bowel loops and suggested a smoothly tapering stricture in the distal ileum. Small-bowel barium enema showed two smooth strictures in the small intestine with proximal hold-up (Figure 1). At laparotomy two strictures were detected in the proximal ileum and the surgeon felt the appearances were 'typical of Crohn's with fat wrapping and skip lesions'. Histological examination was

Departments of ¹Gastroenterology and ²Pathology, Northern General Hospital, Herries Road, Sheffield S5 7AU, UK

Correspondence to: Hal Spencer

E-mail: happyhal@cheerful.com