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

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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

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Figure 1 Small-bowel barium enema showing two discrete smooth strictures with proximal hold-up

to prove otherwise. There were three well demarcated ulcerated areas causing separate stenoses. The ulcers were superficial, affecting the mucosa and submucosa, and did not extend to the underlying tissues (Figure 2). A chronic

mixed inflammatory cell infiltrate containing some eosinophils was present. The lesions were typical of cryptogenic multifocal ulcerous stenosing enteritis (CMUSE).¹ Giant cell granulomas and transmural ulceration were absent, making Crohn's disease unlikely (at a histology meeting none of three pathologists thought the slides consistent with Crohn's), whilst the absence of villous atrophy and malabsorption excluded chronic ulcerative non-granulomatous jejunitis.² Review of the small-bowel barium study confirmed unusual discrete strictures in the jejunum and proximal ileum which were not typical of Crohn's. Damage induced by non-steroidal anti-inflammatory drugs was offered as an alternative radiological explanation, but apart from an occasional aspirin our patient had not taken such agents for many years.

Postoperatively the patient's haemoglobin became normal though her ESR remained raised. Vasculitis due to deficiency of complement component C2 has been suggested as a possible cause of CMUSE,³ but in our patient C2 levels were normal. C3 and C4 levels were also normal. Autoantibodies were all negative.

COMMENT

CMUSE, first described in 1964 by Deray *et al.*,³ is an unusual condition, with fewer than twenty cases reported.⁴ Most cases present with repeated bouts of small-intestine

Figure 2 Haematoxylin-eosin stain showing inflammation limited to the mucosa and submucosa (double arrow) and not affecting the muscularis propria. The mucosal surface demonstrates a well-demarcated ulcer (block arrow)

obstruction. Multiple strictures (2–25) are often present, with frequent relapse following resection. Steroid responsiveness is typical but steroid dependence often occurs. Limited experience with immunosuppressants (azathioprine and methotrexate) suggests lack of benefit.⁴ Weight loss was a feature in about half of the cases described and systemic symptoms such as fever and arthralgia may also be present. Iron deficiency anaemia has not, we think, been previously described as the presenting complaint.

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A tense case— Carney's triad

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A rare differential diagnosis for secondary hypertension is Carney's triad—extra-adrenal paraganglioma, gastric leiomyosarcoma, pulmonary chondroma.¹

CASE HISTORY

A man aged 46 with known refractory hypertension was referred for investigation after a year of occipital headaches on exertion and six months of increasing lethargy and fatigue. Associated symptoms included drenching night sweats, paraesthesiae in his fingers, cold peripheries, a decreasing libido, and weight loss of 7 kg over the past four months. Current treatment was felodipine 10 mg and lisinopril 10 mg daily.

Blood pressure on admission was 162/102 mmHg; routine blood indices were all normal apart from mildly

deranged liver function tests. 24-hour urinary excretion of catecholamines and their metabolites was measured by high-performance liquid chromatography with solid phase extraction while the patient was on the original doses of felodipine and lisinopril. Norepinephrine was 20 000 nmol/24 h (normal range 0–500), epinephrine 24 nmol/24 h (0–100), normetanephrine 25.2 μ mol/24 h (0.1–3.3), dopamine 5480 nmol/24 h (0–3000) and vanillylmandelic acid 114 μ mol/24 h (<35). Chest radiography revealed a 5 cm calcific lesion in the left upper zone consistent with a pulmonary chondroma (Figure 1). This pulmonary lesion was not a new finding. It had been detected at age 26 when the patient had a routine chest radiograph after a road traffic accident in Italy; at that time he had declined further investigation. Ultrasound of the abdomen showed an enlarged fatty liver; the left kidney was mildly hydro-nephrotic but no masses were seen in the upper pole of either kidney. CT revealed a left-sided 7.5 cm para-aortic mass consistent with an extra-adrenal paraganglioma (Figure 2). An iodine-123-meta-iodobenzylguanidine (MIBG) scan was negative, possibly because labetalol had been prescribed earlier and intravenous phenoxybenzamine had been given in preparation for surgery. An attempt was then made at resecting the tumour. Frozen and paraffin sections confirmed that it was a pheochromocytoma with lymph node metastasis. Unfortunately, postoperative CT revealed residual tumour not amenable to further surgery. Endoscopy was performed to exclude gastric leiomyosarcoma. No such lesion was seen and biopsies were negative. Postoperatively, the urinary catecholamine excretion did not become normal but the patient is at present symptom-free on a regimen of sandostatin 50 μ g and phenoxybenzamine 10 mg twice daily and propranolol 40 mg, lisinopril 20 mg and bendrofluazide 2.5 mg daily.

Figure 1 Chest radiograph showing pulmonary chondroma in left upper zone

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