Case report

Unusual cranial and abdominal computed tomographic (CT) scan appearances in a case of systemic lupus erythematosus (SLE)

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SUMMARY A 35 year old woman presented with headache and fever. Computed brain tomography showed diffuse low attenuation in the cerebral white matter. Several months later, serological tests for systemic lupus erythematosus (SLE) became positive. In spite of immunosuppressive therapy she relapsed after six months of treatment, presenting with abdominal symptoms and signs. On this occasion an abdominal CT scan showed distended and oedematous loops of bowel attributed to an underlying vasculitis. This case illustrates novel CT scan appearances in two systems involved in SLE.

Key words: leucoencephalopathy, acute abdomen.

Computed tomographic (CT) scanning has contributed to many aspects of medicine, notably neurology. The case of SLE described here illustrates the radiographic diagnosis of both an unusual neurological complication of this disease and of extensive bowel wall oedema attributed to a lupus vasculitis.

Case report

A 35 year old woman of Yemeni origin presented with a generalised raised erythematous rash, polyarthropathy, meningism, and photophobia. There was no significant drug or past medical history. Examination showed a conscious pyrexial patient with slight memory impairment. The sclerae were injected and the lids puffy. Tender cervical lymphadenopathy was present. There was no neurological abnormality except a haemorrhage adjacent to the left optic disc. The remaining examination was normal apart from traces of blood and protein on urine analysis.

A lumbar puncture showed clear sterile fluid under normal pressure, with normal biochemistry and cells. An unenhanced CT brain scan (Fig. 1) showed diffuse, uniform low attenuation in the white matter (mean (SD) 22 (2.5) Hounsfield units (HU)), normal white matter 30 (2.5).1 There was no change after contrast enhancement. The electroencephalogram showed non-specific changes (increased random theta activity bilaterally with widespread high amplitude delta waves on hyperventilation), indicating a diffuse abnormality. Blood tests showed: erythrocyte sedimentation rate (ESR) 120 mm/h; normal haemoglobin (Hb) and white blood cell count (WBC); mild thrombocytosis (401 x 10^9/l), normal renal and hepatic function; a total haemolytic complement level of <20% (normal range (NR) 60–140); C4 34% (NR 60–140); C3 50% (NR 75–125); repeatedly negative antinuclear factor (ANF); negative syphilis serology (Venereal Disease Research Laboratory test (VDRL)), monospot, Paul-Bunnell reaction, mitochondrial smooth muscle, and gastric parietal cell antibodies; normal serum C reactive protein concentration, antistreptolysin O, toxoplasma, and leptospiral titres. Cryoglobulins were not detected.

A conservative approach was adopted and there was spontaneous improvement, the ESR falling to...
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60 mm/h. During follow up she continued to complain of mild headache and sore eyes. The serum complement factors remained low, and a further small flame haemorrhage appeared near the right optic disc and then disappeared. The traces of blood and protein in the early urine samples disappeared.

Four months later she relapsed with fever, dry mouth, and arthralgia. The ANF was now strongly positive (titre >320) and the DNA binding was 30%. The Hb had fallen to 9.1 g/dl (91 g/l) and the WBC to 3.3×10^9/l. Her cranial CT scan was essentially unchanged. A diagnosis of SLE was made, she was started on oral prednisolone (60 mg/day), and there was immediate symptomatic improvement. Her temperature became normal and the ESR fell to 28 mm/h. The Hb and WBC returned to normal.

She became Cushingoid, and the dose of prednisolone was gradually reduced to 15 mg/day. Although her headaches improved, the complement levels remained low and she complained of arthralgia and dyspepsia. Azathioprine (150 mg/day) was added.

Six months after her second hospital admission she began to complain of severe abdominal pain typical of small bowel colic. She vomited and passed loose watery stool. On examination she was distressed but afebrile, with a distended tender abdomen. The ESR was 52 mm/h. Plain abdominal radiographs showed small bowel fluid levels and suggested some ascites. Bacterial peritonitis was suspected. A diagnostic aspirate of ascitic fluid showed LE cells containing phagocytosed material (Fig. 2). Rapid abdominal distension followed, associated with oliguria and haematuria. The serum creatinine rose to a peak of 400 μmol/l. An abdominal CT scan showed little ascites but distended, oedematous loops of small and large bowel (Fig. 3). Because of the tendency for patients with SLE to develop venous thromboses, portal or mesenteric vein thrombosis was considered. The former seemed unlikely in view of a normal sized spleen shown by
CT; subsequent mesenteric angiography was normal. Small vessel lupus vasculitis involving bowel and kidneys was diagnosed, and the dose of steroids increased to prednisolone 100 mg/day. Her condition rapidly improved. Renal function returned to normal. A repeat CT examination showed complete resolution of the abdominal appearances (Fig. 4) but no definite regression of the original brain abnormality.

Discussion

Late seroconversion of the ANF is recognised in SLE such that the ANF can become positive up to 14 years after presentation with pathology retrospectively attributed to the disease.2

The cranial CT appearances could be described as 'leucoencephalopathy'. We are unable to find a report of such CT appearances in SLE. where the common abnormalities are 'perisulcal atrophy' and infarction.3 Clinically, focal infarction, epilepsy, and psychiatric disturbance are common neurological features in this condition.4 A few cases of progressive multifocal leucoencephalopathy have been observed in SLE,5 6 but this serious and usually fatal condition differs both clinically and radiologically from the present case.

Such diffuse and relatively uniform CT changes confined to the white matter would be unusual in the viral, postinfective, and primary demyelinating encephalitides.7 Several congenital enzyme defects, such as metachromatic leucodystrophy, can present in adult life with a demyelinating encephalopathy, but these usually progress to obvious brain atrophy.8 Diffuse white matter low attenuation is a recognised feature of hypoxic brain injury in both infants and in adults.9-11 and of microangiopathies of various aetiologies: granulomatous angiitis,12 irradiation,13 and subcortical arteriosclerotic encephalopathy.14

The white matter attenuation values of between 20 and 26 HU indicate a reduction in tissue specific gravity.15 These levels are consistent with a 5–8% increase in tissue water but are not sufficiently low to indicate major loss of tissue or conversion of myelin to neutral fats,16 which in any case would be unlikely in the absence of major neurological deficit or of progression to cortical atrophy with ventricular dilatation. The underlying pathological process was probably oedema secondary to a lupus microangiopathy: a vasculitic process selectively involving smaller arteries could produce predominantly white matter changes by affecting mainly the long corticomedullary arteries, so sparing the cortex.

Even after treatment with immunosuppressive therapy with two drugs there was little improvement in the cranial CT appearances. In the absence of major neurological symptoms and signs, however, increase of the doses above those required to control headache and abdominal symptoms was not felt to be justified.

Gastrointestinal manifestations of SLE were first described by Osler.17 Almost any cause of abdominal pain may be simulated, often resulting in misdiagnosis and unnecessary laparotomy.18-20 More recently SLE has become a recognised cause of 'medical peritonitis',21 but the concern has always remained that these patients are more liable to develop a true bacterial peritonitis,22 particularly if receiving immunosuppressive therapy as in this patient. The underlying cause is a lupus vasculitis or venulitis involving the intestinal mucosa and submucosa.18 23 Conventional contrast radiographs of the gut usually show thickened, oedematous loops of bowel,21 24 but we are unaware of any previous reports of abdominal CT scan appearances, the pathophysiology of which was probably the same as in the brain. The importance of abdominal paracentesis should be stressed as this not only excludes the possibility of bacterial peritonitis, but the presence of LE cells may establish the diagnosis of lupus vasculitis.

We conclude that SLE can be complicated by a widespread abnormality of the brain white matter with marked radiological changes but few neurological signs, and that abdominal SLE can manifest as marked bowel oedema on CT scan.

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References

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