Neurogenic bladder due to peripheral neuropathy and a visual disturbance in an elderly man with systemic lupus erythematosus

Takashi Wada, Hitoshi Yokoyama, Kenzo Ikeda, Naohisa Tomosugi, Kenichi Kobayashi

Abstract

A 63 year old man with central nervous system lupus with a neurogenic bladder and visual disturbance is described. The diagnosis of neurogenic bladder, attributed to peripheral neuropathy, was made on the basis of cystometrography and clinical symptoms. A brain magnetic resonance imaging scan showed gliosis along the cerebral vessels and the optic nerve. This case shows that systemic lupus erythematosus can be accompanied by a peripheral neurogenic bladder and visual disturbance, and that these symptoms may not improve despite the amelioration of other lupus symptoms on treatment with steroids.

It is rare for both the bladder and optic nerve to be affected in systemic lupus erythematosus (SLE).¹ Involvement of the bladder may be manifested by lupus cystitis and neurogenic bladder, which may be caused by an upper motor neurone disturbance. The latter is part of the myelopathy of the central nervous system as a result of lupus.^{2,3} Central nervous system lupus may also produce visual and neurological symptoms resembling multiple sclerosis.⁴ We describe here a patient who developed simultaneously a neurogenic bladder caused by a lower motor neurone disturbance and visual disturbance. This is believed to be the first report of such a case.

Case report

In June 1988 a 63 year old Japanese man suddenly developed urinary incontinence with severe general malaise and low grade fever. He was admitted to Kanazawa University Hospital for evaluation on 1 September 1988.

A neurological examination showed hyperactive deep tendon reflexes of the lower legs, and a positive bilateral Babinski reflex. There was no evidence of sensory neuropathy. Examination of the cranial nerves showed right lower medial defects in both visual fields. The patient was unaware of any visual disorder. There was no papilloedema. Urine analysis showed microscopic haematuria and proteinuria of 0.5-1.0 g/day; the urine sediment contained some granular casts. The packed cell volume was 0.28. The white cell count was $3.1 \times 10^9/l$ (neutrophils 58%, lymphocytes 20%). The serum protein was 60 g/l (albumin 25 g/l, γ globulin 12 g/l). Serological testing was negative for C reactive protein (1.0 mg/l) and the erythrocyte sedimentation rate was 33 mm/h. Rheumatoid factor was 113 U/ml, lupus erythematosus test

positive, and antinuclear antibodies positive at a titre of 1/2520. Tests were negative for antibodies to ribonucleoprotein, Sm, SS-A, SS-B, Scl-70, and Jo-1. Unclassified antibody to extractable nuclear antigens was positive. Antibodies to DNA were 600 U/ml (normal <20). Hypergammaglobulinaemia was present: IgG 23.68 g/l (normal 13.25-17.73), IgM 2.14 g/l (normal 0.65-1.67), and IgA 2.33 g/l (normal 1.40-3.16). Components of complements showed low titres; C3 210 mg/l (normal 520-820), C4 90 mg/l (normal 180-420), and CH50 14 U/ml (normal 32-47). Lumbar puncture showed a clear, colourless, acellular cerebrospinal fluid. Analysis of this cerebrospinal fluid showed protein 370 mg/l; IgG 69 mg/l (normal 0-50); CH50 less than 6 U/ml, and antinuclear antibody titre 1/20.

This patient was diagnosed as having SLE based on the revised criteria for its classification.⁵ He did not report any family history of collagen diseases. A renal biopsy specimen was taken on 19 September 1988. Segmental mesangial proliferation, necrosis, haematoxyphil bodies, and 'wire-loop' lesions were seen in the

Figure 1 (A) Initial brain magnetic resonance imaging scan before treatment, T2 weighted image. (A) There are high intensity lesions in the deep white matter and basal ganglia (arrow). (B) High intensity lesion is seen in the right postbulbar region of the optic nerve (arrow).

Division of
Nephrology and
Clinical Immunology,
First Department of
Internal Medicine,
School of Medicine,
Kanazawa University,
Kanazawa,
Japan

T Wada
K Ikeda
H Yokoyama
K Kobayashi

Division of Laboratory,
School of Allied
Medical Professions,
Kanazawa University,
Kanazawa,
Japan
N Tomosugi

Correspondence to:
Dr Takashi Wada,
First Department of
Internal Medicine,
School of Medicine,
Kanazawa University,
13-1 Takara-machi,
Kanazawa,
Japan.

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glomeruli, which we classified as lupus nephritis IIIB according to the WHO classification. A magnetic resonance imaging scan of his brain showed no relevant findings in T1 weighted images. There were high intensity lesions corresponding to the bilateral deep white matter, basal ganglia (fig 1A), and the right postbulbar region of the optic nerve in T2 weighted images (fig 1B). These findings have previously indicated gliosis^{6 7} distributed along the vessels. Cystometrography showed an autonomous neurogenic bladder disturbed by peripheral motor neurones. The patient did not have any history of lumbar injury or of an abdominal operation. Rectal examination revealed mild benign prostatic hypertrophy. A magnetic resonance imaging scan of the lumbar region showed no evidence of gliosis in the spinal cord nor deformity of the lumbar vertebrae. Treatment was begun with prednisolone, 40 mg daily by mouth, and an intravenous pulse of methylprednisolone, 1000 mg daily for three days. The clinical and serological activities of the SLE were reduced after this treatment, with a titre of 1/320 for antinuclear antibodies, 8 U/ml for

antibodies to DNA, and 28 U/ml for antibodies to CH50. There was no proteinuria. A follow up magnetic resonance imaging scan of his brain performed 18 months later showed an increase in the size and number of high intensity lesions, indicating that the gliosis rapidly increased. This differs from the clinical course of arteriosclerosis (Fig 2A and B). The patient could urinate 50 ml voluntarily but cystometrography showed no appreciable improvement. Pyramidal tract signs and the visual disturbance also did not improve despite the treatment with prednisolone and methylprednisolone.

Discussion

Neurological disorders occur in about 10–20% of patients with SLE^{1 8} with convulsions (9.1%) and apoplexy (4.3%) being relatively common. The lower urinary tract is affected in only 1% of patients with SLE, and a definite diagnosis of neurogenic bladder has been reported in only three patients, who all had central nervous system lupus with an upper neurone disorder presenting as a symptom of transverse myelopathy, as shown in the table.^{2 3} The neurogenic bladder in our patient was a result of peripheral neuropathy as determined by the cystometrographic observations and clinical symptoms. Its pathogenesis was uncertain. The patient had no history of lumbar injury or operation. It should be emphasised that he had an acute onset of neurogenic bladder concomitant with an increase in SLE activity. The vasculitis of SLE was probably responsible for this neuropathy, though other causes cannot be excluded.

The visual field is rarely affected in SLE, with only eight patients describing conditions such as optic neuritis.⁴ However, none of these showed optic lesions on ordinary radiographs, and no definite diagnosis of optic neuritis was made before death. In our patient, we detected high intensity lesions in the optic nerve which contributed to the clinical visual defects, as shown by a T2 weighted image of the brain by magnetic resonance imaging. Magnetic resonance imaging was also useful in diagnosing central nervous system lupus in this patient because similar lesions were observed along the vessels in the bilateral deep white matter and basal ganglia.^{6 7} Central nervous system lupus should be differentiated from multiple sclerosis in which optic nerve involvement, known as Devic's syndrome, is common. The distribution of the gliosis detected by magnetic resonance imaging could aid the differential diagnosis of these two diseases: in multiple sclerosis it tends to be limited to the white matter of the cerebrum and not present along the vessels.^{6 7 9} In this patient low levels of complement in the cerebrospinal fluid also supported the diagnosis of central nervous system lupus.^{1 10}

In this instance the diagnosis of central nervous system lupus was made, unusually, in an elderly man. Catoggio *et al* also reported that elderly patients with lupus have shown such unusual manifestations as cutaneous, neuropsychiatric, or pulmonary disorders.¹¹ Although the pathogenesis of SLE in elderly patients is

Figure 2 Follow up brain scan by magnetic resonance imaging after eighteen months. T2 weighted image. (A) An increased number of high intensity lesions are seen in the deep white matter and basal ganglia (arrows). (B) A high intensity lesion is seen in the right postbulbar region of the optic nerve (arrow).

Published reports of neurogenic bladder in patients with SLE

Age/sex	Central nervous system lupus	Lesions	Treatment*	Response	Reference
48/M	+	Upper neurone	P	None	Sugiyama <i>et al</i> ³
18/F	+	Upper neurone	P, M	None	Sugiyama <i>et al</i> ³
(?)	+	Unclear	P, C	None	McCune <i>et al</i> ²
63/M	+	Lower neurone	P, M	None	This paper

*P=prednisolone; M=methylprednisolone pulse treatment; C=cyclophosphamide.

unclear, there are immunological differences between young and elderly patients, including age related changes in T cell functions,¹² and a high frequency of HLA-B8 and DR3 expression in those patients with SLE showing a later onset of disease.¹³ Such immunological changes may be partially responsible for the unusual manifestations of SLE in elderly patients.

The neurogenic bladder and visual disturbance of our patient did not improve although SLE activity was reduced by treatment with prednisolone given by mouth and pulses of methylprednisolone given intravenously. Previous reports of neurogenic bladder as a result of central nervous system lupus have also shown a poor response to treatment.²⁻³ This may be because the optic nerves and brain lesions develop degenerative changes, as detected by magnetic resonance imaging scans. Earlier diagnosis and treatment may be important in achieving an improvement of these atypical symptoms of lupus vasculitis.

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