Multiple Sclerosis and Systemic Lupus Erythematosus in a 10-Year-Old Girl

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Systemic lupus erythematous (SLE) is uncommon in children and multiple sclerosis (MS) is extremely rare. Both the diseases have a chronic, relapsing course and very rarely they have occurred in the same individual^) when a common underlying etiology has been suggested(2). Both SLE and MS are associated with HLA-DR2 allele and have occurred in twins of successive generations(3). We describe co-occurrence of MS and SLE in a 10 year old girl who had the first neurological episode at the age of 5 years. To the best of our knowledge, this is the first description of MS and SLE occurring together in a child.

Case Report

A 10-year-old girl was first hospitalized at the age of 5 years when she had presented with urinary retention. She completely recovered over a period of 10 days but one month later presented with quadriparesis with upper motor neuron signs. She was admitted with the diagnosis of myelopathy with cervical localization. This time also she recovered completely over a period of one month without any specific therapy and remained well till April 1992 when she came to us with acute renal failure requiring peritoneal dialysis. Investigations snowed a strongly positive Fluorescent Antinuclear Antibody (FANA) test. There was no history of malar or other rashes, photosensitivity, joint involvement or oral ulcers. Renal functions normalized and urinary abnormalities disappeared on oral prednisolone therapy over a period of 2 months. She was again hospitalized in September 1992 with a complaint of sudden loss of vision in both the eyes. Examination revealed bilateral optic neuritis. No other neurological abnormality was present. In view of three separate episodes of neurodeficits involving different sites of the central nervous system, she was diagnosed to have clinically definite relapsing remitting multiple sclerosis(4). Two days after admission, she had one episode of generalized tonic-clonic convulsion, which did not recur after she was put on dilantin. There was gradual improvement in vision on steroid therapy. She was able to read fine prints by 18 days of therapy and was discharged from the hospital. She was attending school and leading a normal life till March 1993 when she developed progressive weakness of both the lower limbs and urinary incontinence. She also gave a history of rash on the cheeks and photosensitivity. Physical examination confirmed myelopathy with motor and sensory deficits below D_5 and a fixed erythematous flat rash over the cheeks and bridge of the nose. A diagnosis of SLE was made as she satisfied the suggested criteria(5). She was started on 2 mg/kg/day of oral prednisolone which she received for 7 days. Gradual recovery of muscle power was noted in both the lower limbs and partial bladder control was achieved. She was

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discharged after 1 week on oral prednisolone, which was tapered off over a period of 3 weeks.

Renal biopsy done after her recovery from acute renal failure revealed tubulo-interstitial disease with normal glomeruli *(Fig. 1)*. As has been mentioned earlier FANA was highly positive (+3) and C_3 was only 40 mg/dl. The C_3 level returned to normal during follow up. Unfortunately immunofluorescent study of the renal biopsy specimen could not be done. Urine showed mild proteinuria (albumin 1+) and cellular casts. FANA remained positive in all the subsequent admissions and antibodies to ds-DNA were also positive. Investigations done during the hospital admission for optic neuritis showed oligoclonal bands in cerebrospinal fluid (CSF), abnormal brain stem evoked response audiometry (BERA) and visual evoked responses suggesting demyelination. CT scan of head was, however, normal. Magnetic resonance imaging (MRI) scan was not done as the parents of the child did not agree for the test. CSF examination during the last admission showed 10 cells, all lymphocytes. Glucose was 60 mg/dl (blood glucose 86 mg/dl) and the protein level was 80 mg/dl. There was increase in the gamma-globulin fraction and oligoclonal bands were present. Brain stem and visual evoked responses were again abnormal showing prolonged central conduction.

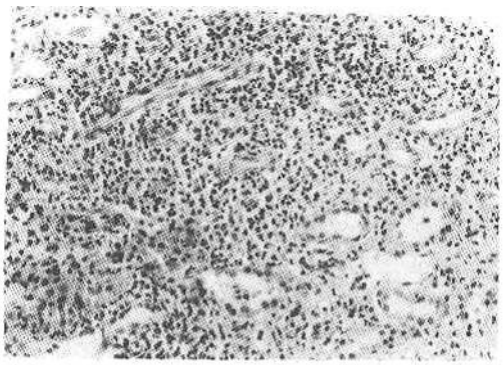

Fig. 1. Photomicrograph of renal biopsy specimen showing dense lymphomononuclear cellular infiltrate in the interstitium, tubular damage and normal glomeruli (H&E ×275).

She gradually regained her power in lower limbs and started attending school. However, the recovery was incomplete. She died at home approximately seven months after her last hospital admission. The cause of death is not known and autopsy was not performed.

Discussion

Multiple sclerosis is rare in childhood. In a population based retrospective study(6), 125 out of 4632 patients with MS (2.7%) had initial manifestations before 16 years of age while 8 patients were symptomatic before 11 years. Bye *et al.(7)* reported 5 cases of childhood MS; their youngest case was a 3 year old girl. Childhood SLE is also an uncommon disease. Only 3.5% of all SLE occur before 10 years of age(8) and the literature is silent on the exact incidence of neurological manifestations in this group of patients. Multiple sclerosis has been associated with SLE occasionally(1,9,10). All reported cases were adults.

Neurological dysfunction antedating systemic and renal manifestations is not commonly seen in SLE; SLE myelopathy is extremely rare. April *et al.(11)* on reviewing the literature could find only 27 cases (age range 11-62 years) of myelopathy in SLE. The causes of lupus myelopathy were vasculitis with myelomalacia, cord necrosis and intradural and extradural hemorrhage. Lupus myelopathy progresses rapidly with no functional recovery(11). There lapsing-remitting course of spinal cord involvement, optic neuritis and the initial presentation with only bladder involvement syn-

thesizes in diagnosis of MS. This diagnosis is further supported by the oligoclonal bands in CSF and abnormal evoked responses with delayed central conduction. Our patient, therefore, probably had both multiple sclerosis and SLE.

Is there any relationship between these two diseases? Both MS and SLE are associated with HLA-DR2 allele(3). Also, they have occurred in twins in successive generations(3) and in two generations of the same family(12). Hence both these diseases may be caused by a common immunogenetic mechanism. A common etiology has been suggested as increased titers of measles antibodies have been demonstrated in both the diseases. Other similarities include defective natural killer cell function(13) and frequent occurrence of antibodies to glycosphingolipids(14).

Acute anuric renal failure is also rare in SLE. Interstitial nephritis with normal glomeruli on histopathology is rarer still. There are only 8 well recorded cases of SLE with predominant tubulointerstitial nephritis(15) and acute renal failure occurred in 5 of them. Other causes of interstitial nephritis *(e.g.,* use of non-steroidal anti-inflammatory drugs) were ruled out on history in our patient.

Our patient, therefore, had an extremely rare combination of MS and SLE in the first decade of her life and a very unusual finding of SLE on renal histopathology.

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