DEMYELINATING DISORDERS

CLINICAL FEATURES OF CHILDHOOD MULTIPLE SCLEROSIS

The clinical and paraclinical features of multiple sclerosis (MS) were reviewed in a study of 36 of 890 (4%) MS patients whose symptoms began before 16 years of age and who were followed at Dokuz Eylul University, Izmir, Turkey. Four cases were lost to follow-up. The average age at onset of MS in children was 12.9 years (range, 8-16 years). Girls (23) were affected more often than boys (9), with a sex ratio of 2.5:1. The sex ratio in the total MS population was 1.47:1, and the difference was highly significant (P<0.0001). Diplopia and sensory disturbances were the most common initial symptoms, occurring in 28% of cases, and a single symptom was noted at onset in 18/32 (56%) patients. Visual impairment due to optic neuritis occurred at onset in 17%. In patients with a monosymptomatic onset, definite MS developed 6 years later. In those with multiple symptoms at onset, MS was diagnosed after 14 years. A non-specific infection, usually URI, occurred in the 2 weeks before onset of MS in the majority of cases. Oligoclonal band was positive in 91% of patients. MRI evidence of white matter MS plaques was found in 96%, and abnormal visual evoked potentials in 65%. SEP and BAEP abnormalities were recorded in 59% and 41%, respectively. A relapsing course occurred in 21 (59%) cases and a secondary progressive course in 11 (30.5%) patients. In 2 patients with an ADEM-like illness at onset, symptoms recurred within 3 months, and more than one relapse occurred later with typical MS symptoms. At the latest follow-up of the 32 patients (average 7.3 years), pyramidal signs were present in 65%, brainstem signs in 58%, cerebellar involvement in 52%, and optic neuritis in 48%. Generalized convulsions occurred in 10%. Two patients (13%) had a positive family history of MS. Two patients had died. (Ozakbas S, Idiman E, Baklan B, Yulug B. Childhood and juvenile onset multiple sclerosis: clinical and paraclinical features. Brain Dev June 2003;25:233-236).

COMMENT. MS in childhood has similar manifestations to that in adults and the prognosis is poor. In contrast to adults, childhood MS may occasionally begin with fever, confusion and convulsions, and coma, a presentation resembling ADEM. In this series, patients with a monosymptomatic onset developed the secondary progressive form of MS more rapidly than those with a polysymptomatic onset.

The lack of awareness of a diagnosis of MS in children is emphasized in a review of MS (Gadoth N. Brain Dev June 2003;25:229-232). An infant or child with acute or subacute neurological disease and MRI evidence of white matter abnormalities will first be considered for neurometabolic disorders, mitochondrial defects, organic and aminoacid urias, leucodystrophies (MLD), infections, ADEM, and vasculopathies. MS is rarely considered in the differential diagnosis despite a frequency in childhood equal to that of metachromatic leukodystrophy. The calculated frequency of childhood MS is 1.35-2.5/100,000, and for infants and young children, it is 0.4-1.4/100,000. In comparison, the estimated frequency of MLD is 2.5/100,000. The possibility of MS should be considered even in infants with neurodegenerative illness.