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MULTIPLE SCLEROSIS IN A SIX-YEAR-OLD BOY*
(CASE REPORT)

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A 6-year-old boy with multiple sclerosis, youngest diagnosed in our hospital, is presented. In addition to the clinical findings, the diagnosis was supported by increased immunglobulin levels, positive IgG index and oligoclonal bands in the cerebrospinal fluid, normal cranial angiography, typical lesions on the cranial computed tomography and magnetic resonance imaging. *Key words: multiple sclerosis, childhood.*

Multiple sclerosis (MS) is a demyelinating disease which affects the white matter of the central nervous system (CNS). MS often occurs in the third decade of life, and is rare under the age of 15. Females are affected more than males. We present the youngest patient with MS ever encountered in our hospital.

Case Report

A six-year-old boy who was in good health until May 1989 was placed on antibiotic therapy with the diagnosis of pneumonia. Four days later he developed ataxic gait and tended to fall to the left.

The patient was a product of an uncomplicated pregnancy and delivery. His growth and development were normal.

On admission to our hospital, he was found to have an ataxic gait, bilateral dysmetria, dysdiadokokinesia, hyperactive deep tendon reflexes (DTR), and bilateral Babinski and clonus signs. He did not display nystagmus or dysarthria. Lumbar puncture revealed normal cerebrospinal fluid (CSF) pressure, 30 cells /

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mm^3, 11 of which were lymphocytes; a protein level of 48 mg / dl; a glucose level of 68 mg / dl; and negative cultures for microorganisms. Electroencephalogram showed a paroxysmal disturbance of electrical activity originating from the right posterior regions of the brain. Cranial computed tomography (CT) showed multiple hypodense areas in the cerebellum. The patient received sulbactam and ampicillin therapy 200 mg / kg qid, as well as dexamethasone 0.6 mg / kg qid orally for 15 days. CT taken on the sixth day of therapy showed that all of the former lesions had disappeared. His symptoms gradually resolved and ceased. He was readmitted eight months later with complaints of nausea, vomiting, gait ataxia, a tendency to fall to the left, holding his head to the left, and swaying to the left. On examination, he had gait ataxia, intentional tremor, uncoordinated movements, hyperactive DTR, bilateral Babinski and clonus signs, head tilt to the left, astenia, hypermetria, dysdiadokokinesia, asinergia and explosive dysarthria. He did not display nystagmus.

Complete blood cell count, urine analysis, hepatic and renal function tests, erythrocyte sedimentation rate, lactic acid, pyruvic acid, ammonia, urine and serum aminoacids were found to be normal; C-reactive protein (CRP), latex, antinuclear antibody (ANA), anti DNA, salmonella, brucella, Paul-Bunnell agglutination tests, vanylmandelic acid in urine were also negative. CSF pressure was normal, with slightly elevated protein (48 mg / dl) and normal glucose levels, with no cells. Bacterial cultures were negative. CT disclosed two hypodense lesions in the posterior fossa with no mass effect or edema. Magnetic resonance imaging (MRI) showed multiple hyperdense lesions of the white matter at T2 weighted imagings which were considered as plaques of multiple sclerosis in the centrum semiovale, pons, mesencephalon, pedunculus cerebelli, around the periventricular region and more often in the left cerebellar hemisphere. IgG in CSF was 7.8 mg / dl (normal range is 1.5 - 5.5 mg / dl). IgG index, which is obtained by measuring CSF IgG / albumin over serum IgG / albumin was 0.8 (ratio greater than 0.77 is generally considered abnormal). Oligoclonal bands in CSF were positive. Brainstem auditory-evoked responses (BAER) showed prolonged central conducting time, while visual-evoked potentials (VEP) were normal.

Dexamethasone therapy was started at a dose of 0.6 mg / kg / day qid, orally. By tapering down the dose, it was stopped within six weeks. On the fifth day of the treatment, cerebellar findings gradually resolved, then subsided.

Discussion

Multiple sclerosis (MS) often occurs in the third decade of life and is rare under the age of 15 and over the age of 50. The incidence of initial manifestations before 16 years of age is 2.7%[1], while the incidence before 10 years of age is 0.3%[2]. The youngest patient documented with MS is a 24-month-old girl[3]. Our case is the youngest MS case ever encountered in our hospital.

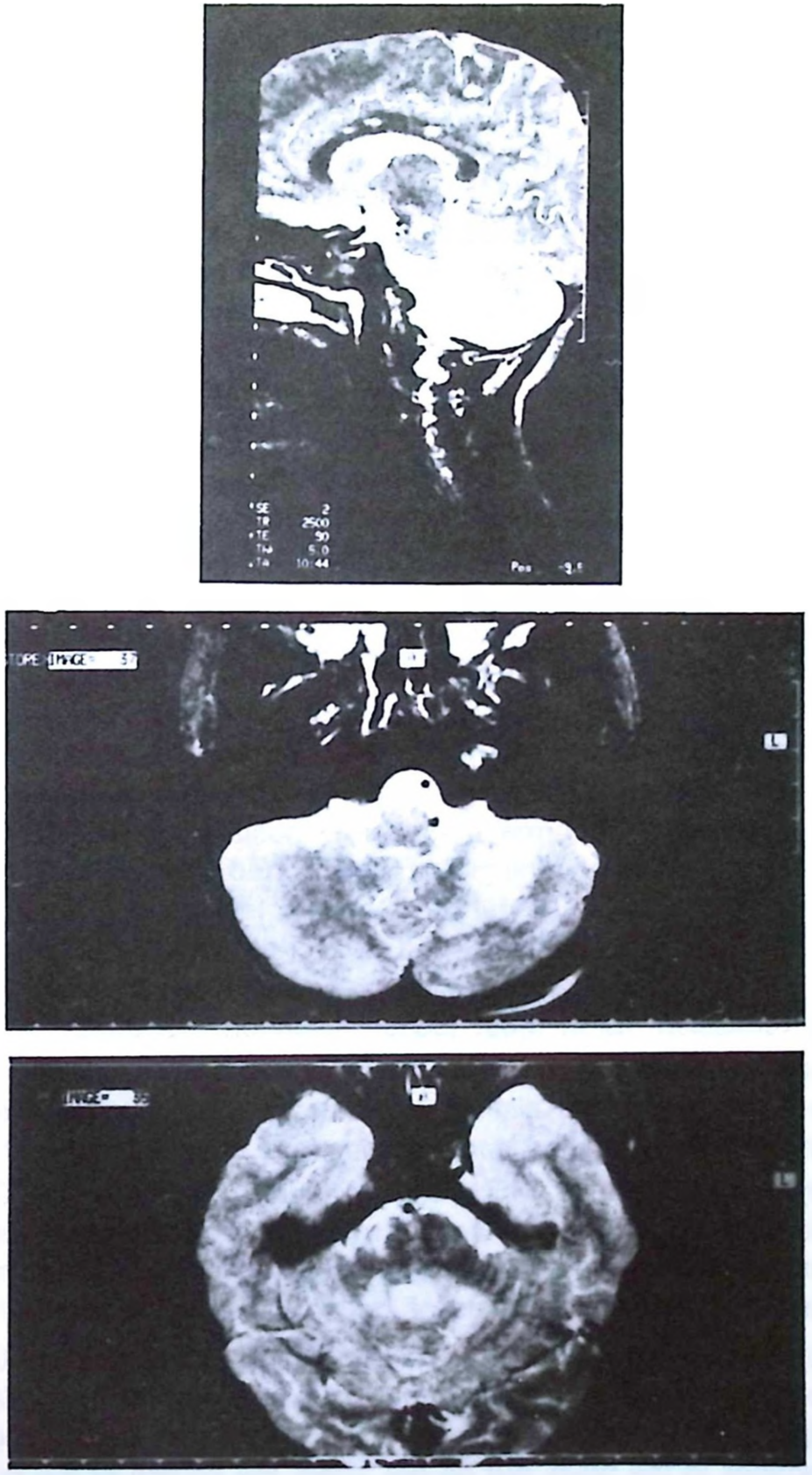

Fig. 1: T2 weighted segments (TR / TE: 2500 / 90 msec) sagittal (a) and transverse (b, c) images.
a) Hyperintense lesions in pons, mesencephalon, corpus callosum and cerebral white matter.
b) Hyperintense lesion in left cerebellar hemisphere.
c) Demyelinating plaque in pons and bilateral dentate nuclei are seen.

The female / male ratio is 2 / 1 in the total MS population and 3 / 1 in the childhood group[1]. On the other hand, a relative overpresentation of boys has been reported in the late-onset forms of MS with poor recovery and a progressive course[2]. Although our patient is male, the disease is neither a late-onset nor a progressive form of MS. This implies that our patient displays a relatively rare property of childhood MS. During both of his hospital admissions, the patient elicited signs of cerebellar dysfunction which denoted involvement of cerebellar hemispheres, partially paleocerebellum and perhaps vermis and / or fasciculus longitudinalis. As initial manifestations, the reported incidences of certain symptoms are as follows: gait problems and ataxia 5-50%, nystagmus 35-75%, dysarthria 6-50%[2,4,5].

Laboratory tests for MS are not specific but have supportive value. Mildly increased CSF protein, leukocyte count of up to 20 / mm^3, and elevation of gammaglobulin to greater than 12% of total protein without any corresponding increase in serum gammaglobulin are notable. IgG index is abnormal in up to 80-90% of patients with MS. Our patient's IgG index was 0.8. A ratio over 0.77 is generally considered abnormal[5]. IgG with an oligoclonal nature, denoting the presence of plasma cells in the plaques, is present at rates of 90% in definite and 30-40% in possible MS cases; nevertheless, it may also be seen in some other inflammatory diseases of the central nervous system (CNS)[5]. By using both BAER and VEP, subclinical lesions are detected in 71% of cases, and the addition of sensory-evoked potentials and blink reflexes contribute an additional 3% of cases[6,7]. Our patient's abnormal IgG index, positive oligoclonal bands, and prolonged central conduction time in BAER supported the diagnosis. Although CT is very helpful, MRI is more useful for detecting lesions in the posterior fossa which are poorly demonstrated by CT and also for detecting silent plaques except on the optic nerve[8,9]. The patient's clinical course was characterized by periods of relapse and remission, as in most (56%) childhood MS attacks.

By Poser's classification, our patient is consistent with a clinically definite MS, as there were two attacks and clinical evidence of two lesions. Those attacks occurred one month apart and each lasted more than 24 hours[10].

Other differential possibilities which cause multiple lesions of the CNS with a course of relapes and remissions include meningovascular syphilis, sarcoidosis, cerebellar abcesses, arteriovenous malformations and vasculitis. In our case these are all excluded on the basis of both clinical and laboratory data.

There is no definite therapy for MS. Adrenocorticotropine hormon (ACTH) and corticosteroids are used in the early phase, especially for the relapsing and remitting type. These medications shorten the duration of attacks and modulate immune reactions in the brain, but do not affect the course of the disease[11,12]. Azathioprine, cyclophosphamide, cyclosporin, lymphatic irradiation and plasmapheresis are being used as a trial[11]. Physical therapy and psychotherapy are also useful in the chronic phase.

We found that our patient's cerebellar findings gradually resolved from the beginning of the fifth day of the oral corticosteroid therapy, which was stopped within six weeks by gradual reduction. Since then the patient has been well and has not had any complaints.

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