Multiple Sclerosis in Childhood: Presenting as Bilateral Optic Neuritis

Radhkrishna H.
Ashok Kumar K.
Srinivashulu C.

Multiple sclerosis is a disease of adolescence and middle age. Though rare, it is being increasingly recognized in younger population. We describe two patients in whom multiple sclerosis was diagnosed before 10 years of age. Both patients had bilateral optic neuritis during the course of their illness with partial recovery; other lesions in the neuraxis recovered reasonably well.

Keywords: Multiple sclerosis, Optic neuritis.

Multiple sclerosis (MS) is being frequently diagnosed in our country and it forms the differential diagnosis of many myelopathies. Patients undergo visual evoked response (VER) tests and cranial magnetic resonance imaging (MRI) scans in search of white matter plaques; though many a time the search is futile. According to the well-established Schumacher’s(1) and Poser’s criteria(2), onset of the symptoms between 10-50 years of age is important. While this is true in most cases, patients outside this age range are increasingly coming to medical attention.

We describe two patients, both of whom had onset of symptoms at less than 10 years of age. The presentation and symptomatology are also different from the usual ‘typical’ cases of multiple sclerosis.

Case Report

Case 1

A 9-year-female child presented with binocular visual impairment, developing over 24 hour period, 4 days back. There was history of right-sided hemiplegia in her 7th year of age, which had improved within 6 months. No imaging was done at that time.

On examination, the visual acuity was limited to perception of light (PL) only in both eyes. Pupils were dilated and reacted sluggishly to light. The optic fundi showed markedly pale discs suggestive of optic atrophy with slightly attenuated vessels. Other cranial nerves and speech were normal. Except mild hyperreflexia on the right side, the rest of the neurological examination was unremarkable.

Visual evoked responses (VER) were grossly abnormal with poor wave morphology, reduced amplitudes and delayed latencies in both the eyes. Cranial MRI scanning showed multiple demyelinating plaques bilaterally, especially in the posterior periventricular white matter and some involving the subcortical ‘U’ fibers.

The optic disc pallor was interpreted as evidence of pre-existing silent optic neuropathy which could have occurred during the previous episode over which the present episode of ‘recurrent’ neuropathy had set in(3).

She was given intravenous methylprednisolone 30 mg/kg for 3 days followed by 11 days of oral prednisolone as per the ‘Optic Neuropathy Treatment Protocol’(4). At review 2 weeks later, the vision had slightly improved to ‘perception of hand movements’ (HM) in both the eyes. The optic disc pallor was persisting. Five months later, she again
presented with recurrence of visual impairment for which she was started on steroids.

Case 2

A 10-year-old male child presented with history of binocular visual impairment since 2 months, which initially started in the left eye and then involved the right eye over a period of 4–5 days. The vision deteriorated over a period of 10–12 days and then remained static. He had no visual complaints earlier. There was a history of quadriparesis with involvement of bladder, 1½ years ago which improved over a period of 6 months. Imaging was not done at that time. There was a history of dog bite 2 years ago for which he received 14 injections of neural antirabies vaccine. The dog did not bite any other person and is still alive.

Examination revealed a binocular impairment of visual acuity limited to counting fingers at 1 m in left eye and at 3 m in the right eye. The left pupil was mid-dilated and sluggishly reacting to light, while the right pupil was normal in size and was reacting well to light. Fundus examination showed moderate optic disc pallor with slight attenuation of vessels on both sides. Other cranial nerves and speech were normal. Motor system examination showed spasticity and mild (grade 4/5) pyramidal weakness in both upper limbs and lower limbs. The deep tendon reflexes were brisk, abdominals were absent and plantars were extensor on both sides. Sensory system examination was normal.

On investigation, the hemogram and other blood parameters were normal. VER were abnormal showing delayed latencies in both eyes with slightly reduced amplitudes in the left eye. A computerized tomographic scan of brain was normal. The patient could not afford MRI scan of brain and the cervical spine.

He was given oral prednisolone 1 mg/kg/day for 14 days. At the time of discharge his vision improved to 6/36 in the left eye and to 6/24 in the right eye. The optic disc pallor improved markedly.

Discussion

Childhood MS is thought to be a rare phenomenon. These two cases illustrate that MS with dissemination in space and time is not uncommon in children. The unusual or ‘atypical’ features noted in these two cases were:

(a) Age of onset below 10 years.

(b) Optic neuritis involving both the eyes in these two cases. Generally, unilateral optic neuritis or retrobulbar neuritis is suggested as a predisposing factor for MS.

(c) In the Indian subcontinent optico-spinal or optico-bulbar forms are commonly seen in adults. Case 1 presented with hemiplegia followed 2 years later by bilateral optic neuritis, whereas Case 2 presented as an opticospinal form, with initial spinal cord involvement, followed 1½ years later by bilateral optic neuritis.

(d) Significant residual visual impairment was noted in both the patients, while the other deficits—paraparesis or hemiparesis, showed good improvement.

The possibility of recurrent ADEM also needs to be considered in children with a clinical picture suggestive of MS. ADEM is generally a monophasic illness characterized by depressed sensorium, seizures and focal deficits. However, occasionally ADEM can be recurrent, which raises the possibility of MS. Recurrences in ADEM generally occur within 6 months after the initial presentation, probably due to premature cessation of steroid therapy and more importantly, the recurrences tend to involve the same site (rather than different sites, as occurs in MS). In our patients, recurrences occurred at intervals longer than six months and involved different sites.
sites. Hence, these cases are more likely to be MS than recurrent ADEM. The VERs in both patients further support the possibility of MS.

In a long term follow up study(5), twenty-five children ranging from 3 to 18 years with an initial diagnosis of Acute Disseminated Encephalomyelitis (ADEM) were followed in the Clinic of Child Neurology for a period of 2 to 8 years. Ten of these children developed Clinically Definite or Laboratory-Supported Definite Multiple Sclerosis during the follow up period. The other 15 children in the study were considered as having Suspected Multiple Sclerosis. Brain magnetic resonance imaging (MRI) performed in the other 15 children disclosed multiple hyperintense lesions on T2-weighted imaging in 13 children; 10 with Definite Multiple Sclerosis and 3 with Suspected Multiple Sclerosis.

Cerebrospinal fluid (CSF) oligoclonal banding was not done in our cases, due to lack of such facility. Earlier studies showed a preferential IgG (Kappa) banding in the CSF to be a sensitive diagnostic index for multiple sclerosis(6).

Finally, it must be emphasized that the diagnosis of MS is purely “Clinical”. While MRI scans and CSF oligoclonal bands support the diagnosis of MS, they are not required in all cases. Patients presenting with more than one attack of clinically definite involvement of white matter should be considered as cases of ‘definite MS’(3). In countries like ours, with scarce diagnostic resources like MRI scanners and assay of oligoclonal bands in the CSF, and the financial constraints of the patients, awaiting results of these investigations (which are required only in clinically doubtful cases to diagnose MS) will only lead to missing the diagnosis. With these considerations, the need for a separate set of criteria formulated to define the course and prognosis of childhood MS seems relevant and timely.

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REFERENCES


