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Case Report

Multiple Sclerosis First Presentation in a Child: Diffuse Motor Demyelinating Polyradiculopathy: Case Report

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Abstract:
Multiple sclerosis is a mostly debilitating chronic demyelinating immune disease of the central nervous system that compromises quality of life within 10-15 years of onset. The clinical picture spectrum varies widely depending on the underlying pathogenesis of demyelination; the macrophage induced demyelination, antibody mediated against myelin proteins, T cell-mediated small vessel vasculitis and subsequent ischemia or cytokine mediated oligodendrocyte death. Axonal injury is limited to 5% of cases. Peripheral nerve demyelination is an exceptionally rare association of multiple sclerosis. We report a 9-year-old who developed bilateral lower limb weakness and loss of truncal support. Electromyography revealed demyelinating diffuse motor polyradiculopathy. He was diagnosed as Guillain-Barré syndrome and received intravenous immunoglobulins with no response. Magnetic resonance imaging of the brain and cervical spine revealed multiple cerebral and cerebellar foci of abnormal signal representing demyelination. He was diagnosed as multiple sclerosis and had a full recovery within 3 weeks from start of management by methylprednisolone. Peripheral demyelinating neuropathy may be the initial presentation of multiple sclerosis in a child.

Level of Evidence of Study: IV (*1*)**.**
Keywords: multiple sclerosis; bilateral lower limb weakness; peripheral nerve demyelinating diffuse motor polyradiculopathy
Abbreviations: EMG: electromyography; MRI: magnetic resonance imaging; MS: multiple sclerosis; NCV: nerve conduction velocity; RIS: radiological isolated syndrome

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Introduction

Multiple sclerosis (MS) is a mostly debilitating chronic demyelinating immune disease of the central nervous system that compromises quality of life within 10-15 years of onset. It is an acquired disease of unknown etiology. Its diagnosis is challenging and its clinical presentation ranges from the clinically silent which is recognized radiologically, i.e. radiological isolated syndrome (RIS), till the florid recurrent demyelinating disease with late axonal damage (*2*). MS initial presentation is often confused with other diseases. The McDonald criteria and scoring system of diagnosis of MS is continuously being revised (*3*). The magnetic resonance imaging (MRI) and other neuroimaging modalities allowed more accurate MS diagnosis and ruling out of other diagnoses (*4*). We report a 9-year-old who developed bilateral lower limb weakness and loss of truncal support. Electromyography revealed demyelinating diffuse motor polyradiculopathy. He was misdiagnosed as Guillain-Barré syndrome and proved to have multiple sclerosis and had a full recovery within 3 weeks from start of management by methylprednisolone. Peripheral demyelinating neuropathy may be the initial presentation of multiple sclerosis in a child.

Case Presentation

A male child 9 years old, presented with history of acute onset with bilateral lower limb weakness and loss of truncal support. His mom reported history of upper respiratory tract infection 3 weeks before the presentation. Urgent electromyography (EMG) and nerve conduction velocity (NCV) showed diffuse motor demyelinating polyradiculopathy, and IV immunoglobulins 2gm/kg were started over 2 days with no response. We retraced the steps. There was no history of march and ascending pattern of weakness, there was complete loss of urinary control since the onset of the illness, and there was no cranial nerve affection. By

examination there was no sensory level, there was bilateral lower limb hypotonia, hyperreflexia and pathological reflexes were present. The brain MRI showed bilateral scattered supratentorial (subcortical, centrum semioval and periventricular), bilateral posterior limb of internal capsule and to a lesser extent cerebellar white matter foci and rounded areas of abnormal signal eliciting low signal in T1, high signal in T2 and FLAIR with no diffusion restriction. There was no blooming in SW1 and no post contrast enhancement. The MRI did not show areas of fresh blood signal intensity and showed normal cervical spine.

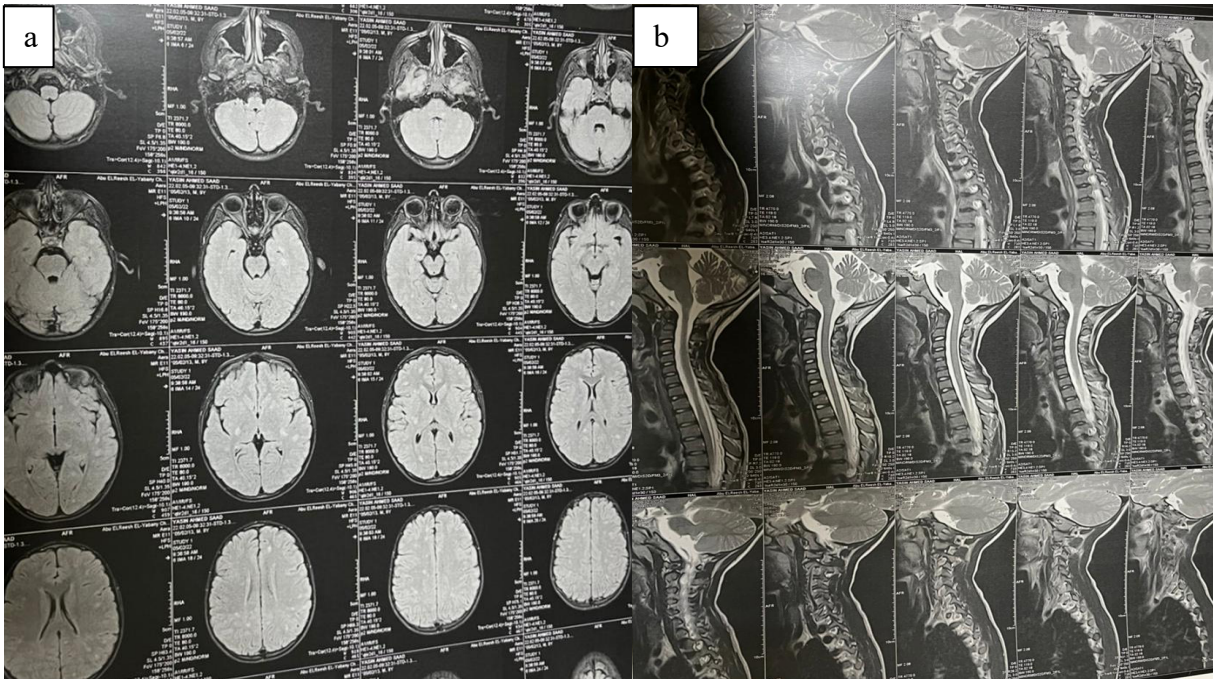

Figure 1. (a) MRI brain, axial cut(*FLAIR*) showed bilateral scattered supratentorial (subcortical, centrum semioval and periventricular), bilateral posterior limb of internal capsule rounded areas of abnormal signal (low signal in T1 and high abnormal in T2 and *FLAIR*) with no restricted diffusion. (b) normal MRI of the cervical spine.

The child was diagnosed provisionally as MS and received IV methylprednisolone 30 mg/kg/d for 5 days then continued on oral prednisolone on 2 mg/kg/day. The patient improved markedly after the second dose then after one week, he started to stand alone unsupported. Within 3 weeks he recovered completely, he walked unsupported and regained urinary control.

Discussion

Initial first presentation of MS is often confused with other diseases as amyotrophic lateral sclerosis, infections as Lyme disease, myasthenia gravis, acute disseminated encephalomyelitis etc. While MS has long been described as exclusively central nervous system disease, this ideology was changed as 5% have axonal peripheral neuropathy.

Yet, peripheral demyelinating neuropathy was reported to be rare, with only 7 cases encountered between 1980 and 2013 among patients at Mayo Clinic (*5*). It is not known if peripheral demyelinating neuropathy is underdiagnosed as EMG and NCV are not part of the regular work up for MS, or it is actually a rare presentation. The spectrum of MS is ever increasing and includes the asymptomatic radiological isolated syndrome (RIS). The promising effect of timely methyl prednisolone in the initial phase of the disease halts and retards the irreversible neurodegeneration. Hence it is of paramount importance to make the diagnosis of MS promptly and timely. While neuroimaging has developed immensely (*6*), the clinical examination and high index of suspicion remains the cornerstone of diagnosis of MS. Timely diagnosis of MS offer the 25% who are refractory to methylprednisolone, other treatment modalities as therapeutic plasma exchange or immunoadsorption to control the neurological damage and reduce disability (*7*).

Conclusion

Peripheral demyelinating neuropathy may be the initial presentation of multiple sclerosis in a child, it might mask the underlying MS. History and examination remain crucial in diagnosis of MS. Children with demyelinating peripheral neuropathy not conforming with the march of disease and typical picture of Guillain-Barré syndrome should be investigated thoroughly and should undergo MRI to exclude MS.

Author Contributions

All authors shared in drafting the manuscript and approved the final manuscript.

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CONFLICT OF INTEREST

The authors declare no conflict of interest in connection with the reported study. Authors declare veracity of information. The datasets generated and/or analyzed for this study are available from the corresponding author upon reasonable request.

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