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A Case Report Of Paediatric Onset Multiple Sclerosis

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Abstract

Multiple sclerosis is a chronic inflammatory demyelinating neurodegenerative autoimmune disorder of the Central nervous system(CNS) of unknown etiology. The peak age of onset is between 20 years to 40 years of age, commonly affecting females; it is common in developed countries. There is less knowledge regarding diagnosis and treatment of Pediatric onset multiple sclerosis(POMS) as compared to adults. Here is a case of Multiple Sclerosis in a 15-year-old girl, it's slow presentation, diagnosis, use of available treatment, and gradual improvement over a few weeks.

Introduction

Multiple sclerosis (MS) is an acquired demyelinating disease of the central nervous system [1]. The etiology of MS is multi-factorial, related to environmental, genetic, and many other factors [2]. The peak age of onset is between 20 years to 40 years of age [3]. It usually affects women more than men [3,4] and is more commonly seen in developed countries compared to developing countries. Pediatric onset multiple sclerosis (POMS) generally occurs before sixteen years of age (maybe sometimes before eighteen years) and makes up about 3-10% of total MS cases [5]. Children generally have a more active disease course than of the disease, with a higher relapse rate. And in neuroimaging POMS shows a greater lesion load

[6]. It is associated with high inflammatory activity. MS in children is usually more frequent and has severe relapses [7]. Anti-CD20 therapies, Rituximab have well-established efficacy in treating immune-mediated neurological and some non-neurological diseases in adults. No disease-modifying drugs have yet been approved for POMS [5]. Some retrospective case series studies have shown the effectiveness of Rituximab, a B cell-depleting anti-CD20 monoclonal antibody [8]. This case report aims to describe POMS in a fifteen-year-old girl who had symptoms of MS for the last 2 years and discuss the brief knowledge of the diagnosis of MS and the treatment options.

Case Presentation

A 15 years old girl from a rural area of a developing nation presented to the outpatient department with complaints of weakness in both upper and lower limbs associated with tremulousness, numbness, and paraesthesia for the last 2 years. She has been treated with repeated courses of multivitamins with little improvement intermittently. There was a history of visual disturbance for 15 days in December 2022 for which no treatment was taken. In April 2023 she suddenly developed visual impairment with near complete loss of vision and was diagnosed with Optic neuritis by ophthalmologists, which readily improved with Steroids (iv Methylprednisolone followed by Tab Prednisolone with tapering dosage). She had no history of muscle weakness, gait disturbance, fever, or urinary Incontinence during the episode. There was no history of Seizure, Loss of consciousness, or headache. Bladder bowel control was normal. Sleep cycle and routine were normal, with above-average school performance. Vaccination was completed according to the IAP schedule. She does not have any known allergies. She attained her menarche at the age of 10 years and had a regular menstrual cycle. Physical examination revealed a girl of weight 80 kg, height 172 cm, BMI 26.8, with all vitals within normal limits during presentation. Blood pressure:

110/90 mm Hg, respiratory rate: 18 breaths per minute.

A complete neurological examination was done. She was conscious oriented to time, place, and person, GCS 15 out of 15, cooperative, with normal intelligence and intact memory, adequate sleep with no obvious speech abnormality. Gait was abnormal in the form of mild spasticity. She was able to comprehend all the commands and was providing all the history by herself. Sense of smell was intact. Extra-ocular muscles were checked for without any abnormal findings. There was no ptosis. The pupil size was 2 mm and reacting well to light and the accommodation reflex was intact. Color vision was normal. No Relative afferent pupillary defect was there. Fundoscopy showed Pale fundus and 6/6 visual acuity with refractory glasses. Sensation in the face and scalp, muscle coordination in the face and neck, hearing and balance, swallowing and the gag reflex, and movement of the tongue were all normal.

Motor examination revealed increased tone across all joints of the upper and lower limbs. The power of muscles of the upper limb across the shoulder and elbow was grade 4 out of 5. Power across the hip, knee, and ankle joint revealed grade 4 out of 5. She was unable to rise from supine posture to sitting upright without using her arms, suggesting weakened abdominal and trunk muscles.

The abdominal reflex was tested to be intact. Plantar was extensor. Deep Tendon Reflex for knee, ankle, and biceps were brisk (3+).

Cerebellar coordination was intact, Romberg sign was absent. There were fine tremors in bilateral upper limbs in both open or closed eyes and aggravated with activity, fatigue, or emotion. Pain, Temperature were intact in dermatomes. tactile localization, tactile discrimination, stereognosis, and graphesthesia were all intact. There were no signs of any nerve root entrapment. Cardiac, pulmonary, and abdominal examinations were unremarkable.

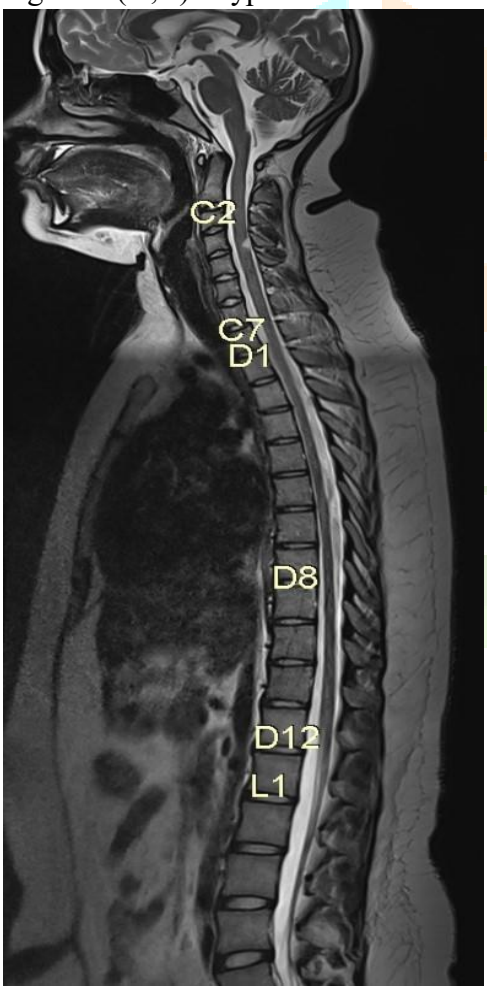
All routine blood tests including Thyroid profile were normal. Ultrasonography showed an incidental finding of a left ovarian cystic lesion. EEG was done and found to be normal. MRI spine showed ill defined short segment hyper-intense lesion involving the cervical cord at C4-C5 disc level causing minimal swelling, suggestive of demyelinating etiology [Figure1 (A,B)]. MRI brain and orbit showed multiple

hyper-intense lesions on flair in the bilateral centrum semiovale, left peri trigonal, bilateral peri-ventricular white matter region, splenium of the corpus callosum region, with focal restricted diffusion in splenium of corpus callosum, and left periventricular white matter lesion[Figure.2]. Both optic nerves were intact. Findings were consistent with Multiple Sclerosis plaque. CSF for oligoclonal band was positive suggestive of Intrathecal IgG synthesis.

Aquaporin 4 NMO antibody and Myelin oligodendrocyte glycoprotein antibody were negative. VEP study showed a right-sided demyelinating type of optic

neuritis.

Figure 1 (A,B): Hyperintense lesions in the c4 and c5 areas of the spinal cord (Arrow mark).



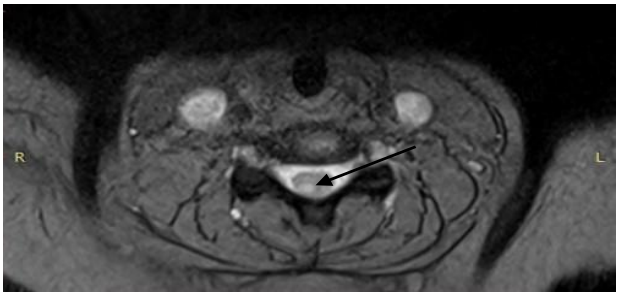
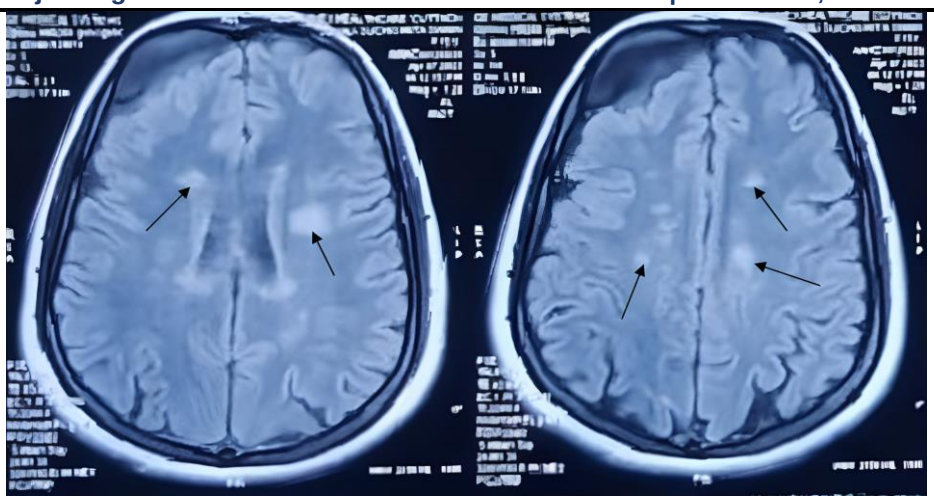


Figure 2: Bilateral multiple hyperintense lesions in in the bilateral centrum semiovale, left peri trigonal, bilateral periventricular white matter region, splenium of the corpus callosum region suggestive of multiple sclerosis plaques. (Arrow marks)



The final diagnosis was made as pediatric-onset multiple sclerosis (POMS).

After the patient's parents' counseling and consent, Rituximab was offered. Before the treatment, Hepatitis B virus Anti HbcAg total antibody and other viral profiles were done which were non-reactive. The first dose of Injection Rituximab 500mg was given by IV infusion, without any adverse reaction. On subsequent follow up child was doing clinically well without any complaints of weakness, numbness, or paresthesia. CD19 cell % decreased to 0.8%(Normal=4.6 to 22.1%), with absolute CD19 cell count 11.2 cells/microL (Normal = 56.6 to 417.4cells/microL). She is now planned for regular follow-ups to track the disease course.

Discussion

Multiple Sclerosis is described as a chronic inflammatory demyelinating neuro-degenerative disorder of the Central nervous system where the myelin sheaths of axons get damaged in the Central Nervous System due to autoimmune processes. In pediatric-onset MS (POMS), the progress is slow as well as atypical, compared to adults. POMS generally occurs before 16 years of age and makes up about 3% to 10% of total MS cases. This affects more girls than boys and the female-to-male ratio varies by age which is 3:1 among adolescent children with the median age group being 12 to 13 years [9]. Obesity is associated with a significantly increased risk of pediatric MS [10]. The efficacy, safety, and tolerability of most of the disease-modifying drugs have not yet been proven. Children generally have a more active disease course

than of the disease, with a higher relapse rate. And in neuroimaging POMS shows a greater lesion load. Children are less likely to develop primary or secondary progressive MS in childhood. More than 95% of pediatric MS patients present with a relapsing-remitting(RR) course, compared with more than 80% of adult patients. Clinical features of POMS and adult MS are very similar; however, children manifest more with optic neuritis, brain stem syndrome, and features of encephalopathy like vomiting, seizure, headache, and/or altered sensorium [11]. McDonald criteria, which is used as diagnostic criteria for adult MS, should not be used as diagnostic criteria in POMS presenting with encephalopathy and multifocal neurological deficit. There is not a single diagnostic test for MS and the diagnosis is usually based on the clinical presentation, supported by neuroimaging like MRI. In some cases, CSF analysis can be done to look for inflammatory markers, oligo-clonal bands, and/or elevated Immunoglobulin G levels. Evoked potential studies can be done to look for a clinically silent lesion in the visual, brainstem, or spinal cord pathways. The inflammatory markers in CSF are present in up to 85% of patients with MS. The IgG index is less sensitive and specific than oligo-clonal bands. T2-weighted MRI among POMS shows more hyperintense lesions in the posterior fossa. Lesions are more reversible on follow-up imaging in children as well as it suggest a better course of recovery. POMS, a highly inflammatory disease, with more frequent relapses compared to adult MS. There has been much advances in the field of diagnosis and treatment of MS in adults but it remains a topic of discussion and research as far as POMS is concerned. Unfortunately, there is no proven or well documented cure for MS. So, treatment is directed towards slowing down the progression of MS, shortening the frequency and duration of relapses alongside managing symptoms [12]. No disease-modifying drugs have been approved for POMS [13]. Few retrospective studies have shown the successful use of Rituximab. In our case, we used Rituximab with encouraging results till now. This case needs to be observed in further serial follow-up.

Conclusions

POMS is a chronic disorder of central nervous system, which is often diagnosed late. In a developing country like India, this case of POMS took almost 2 years to reach an appropriate diagnosis and treatment initiation, because of a lack of awareness about the disease. However, we should acknowledge that POMS is a reality and should be promptly diagnosed and managed, though it needs further study and research.

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