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Case Report

Bilateral ptosis with ophthalmoplegia externa in paediatric onset multiple sclerosis: A case report

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ABSTRACT

Paediatric onset multiple sclerosis (POMS) patients usually present with a diminution of vision pertaining to optic neuritis and experience a more aggressive disease onset with disabling clinical symptoms, a polyfocal presentation at disease onset, and a higher relapse rate early in the disease course. We are reporting a unique presentation of bilateral ptosis with external ophthalmoplegia in a patient of a 17-year-old female patient who came with chief complaint of drooping of lids in both eyes for 3 years. Complete ophthalmic and systemic examination was done. Patient was diagnosed as MS according to the revised Mcdonald criteria and was given Methylprednisolone Pulse therapy. We concluded that acquired bilateral ptosis with ophthalmoplegia externa should be given special attention to rule out systemic associations. Timely diagnosis and early management of multiple sclerosis in the pediatric age group with DMT may reduce relapse frequency and functional disability.

1. Introduction

Paediatric-onset Multiple Sclerosis (POMS) is generally characterized by onset before the age of 18 years. 3 and 10% of patients with MS present under 18 years of age and < 1% under 10 years of age.[1] 84% of adult-onset MS patients present with a relapsing–remitting (RR) course compared to 98% of paediatric-onset MS patients.[2,3] POMS patients generally experience a more aggressive disease onset with disabling clinical symptoms, a polyfocal presentation at disease onset, and a higher relapse rate early in the disease course.[4–6] Patients with POMS usually present with a diminution of vision pertaining to optic neuritis. We are reporting a unique presentation of bilateral ptosis with external ophthalmoplegia in a patient with paediatric-onset multiple sclerosis.

We are reporting a unique case of bilateral ptosis with external ophthalmoplegia in a patient with paediatric-onset multiple sclerosis.

2. Case Report

A 17-year-old female patient came to outpatient department with chief Complaint of drooping of lids in both eyes for 3 years that was gradual in onset, progressive in nature not associated with pain on eye movements. There was no associated history of altered sensorium, fever, headache, vomiting, rashes or head injury. Patient developed dull aching pain in bilateral calf and thighs aggravated with exertion that usually gets relieved with rest. No complaint of dysphagia/ dysarthria/ difficulty in walking/ squatting/ in getting up from sitting.

There was a history of development of bilateral calf pain and weakness in limbs 6 years back for which she did not take any medicine and that was relieved within few weeks.

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There was history of bed wetting till 5 years of age and no history of delayed milestones.

Patient was non vegetarian diet. and had normal bladder bowel habit with normal sleep pattern. There was no history of diabetes mellitus, hypertension or thyroid disease.

2.1. Family history

No of similar complaints.

2.2. Examination

BCVA in Both eyes was 20/20. Bilateral moderate ptosis with restricted ocular movement in all directions with raised eyebrows without head posture.

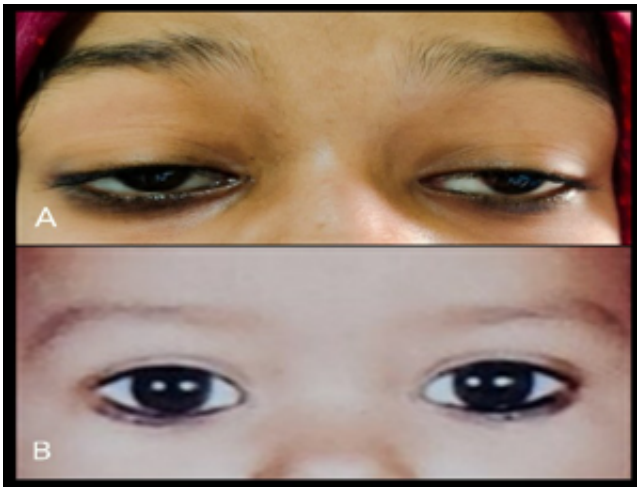

Fig. 1: a: Present picture showing bilateral moderate ptosis with raised brows**. b:** early childhood picture showing normal eyelid position and palpebral fissure

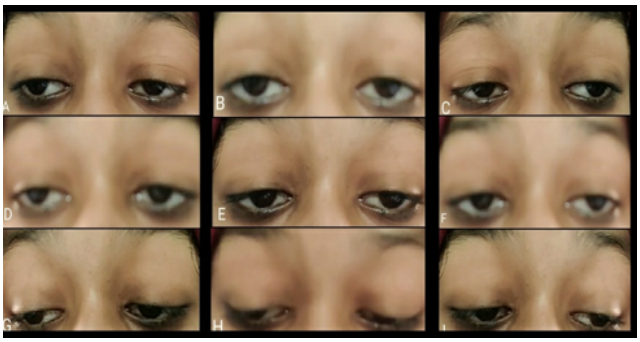

Fig. 2: Restricted Extra-Ocular movements in all 9 Gazes with No change in the position of eyelids on different positions of gazes.

No variation in the amount of ptosis with jaw muscle movement Pupillary Reaction was brisk and sustained in BE. Rest anterior segment examination was normal. Both eyes MRD 1= 1 mm and MRD 2= 6mm. Levator function was poor, with absent lid lag and lagophthalmos. The results of the Ice pack test, Neostigmine test, and Fatigability test were negative. Fundus examination was normal with no signs of optic neuritis. No signs of cerebellar ataxia, or sensorineural hearing loss were noted.

2.3. Investigations

Smooth muscle antibody was Negative (28 IU/ml), Lactate within the normal range (7.3mg/ml) and Acetylcholine receptor antibody was also within the normal range (0.4nmol/L). CSF Examination- normal cytology and biochemistry, with no oligoclonal band (OCB). Anti-NMO antibodies were negative.

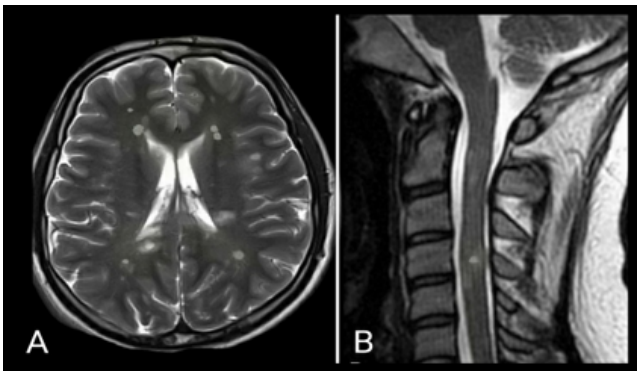

Fig. 3: MRI Brain T2 sequence showinghyperintense lesions in the sub-cortical white matter of the right frontal lobe, the periventricular white matter of bilateral fronto-parieto-occipital lobes. (B) MRI of the Spinal cord showing hyperintense lesion in the C3 region.

According to Polman (2010 revised Mcdonald criteria) diagnosis of multiple sclerosis (MS) was made that requires one of the following findings.[7]

1. ≥ 2 non-encephalopathic, clinical CNS events with presumed inflammatory cause.
2. separated by > 30 days and involving more than one CNS area.
3. One non-encephalopathic episode typical of MS which is associated with MRI findings.

2.4. Treatment

The patient was started on Pulse IV Methylprednisolone 1 Gram per day for three consecutive days and then on oral Prednisolone in tapering dose.

3. Discussion

Paediatric MS has long been an underdiagnosed and undertreated condition. It has distinctive features and the disease course is different than in adult-onset MS.

Patients with POMS usually present with a diminution of vision pertaining to optic neuritis.

Paediatric MS needs to be differentiated from Clinically isolated syndrome (CIS), Acute disseminated encephalomyelitis (ADEM), and Neuromyelitis Optics (NMO).

CIS was ruled out as, it was not the first monofocal/polyfocal, clinical CNS event with presumed inflammatory demyelinating cause with the absence of encephalopathy that cannot be explained by fever. There was a prior clinical history of CNS demyelinating disease with Brain MRI meeting diagnostic criteria for MS

ADEM was ruled out, it was not the first polyfocal, clinical CNS event with presumed inflammatory demyelinating cause with the absence of encephalopathy that cannot be explained by fever.

Absence of typical features on brain MRI during the acute (three-month) phase

1. Diffuse, poorly demarcated, large (> 1–2 cm) lesions involving cerebral white matter.
2. Deep grey matter lesions (e.g., thalamus or basal ganglia) may be present.

NMO was ruled out as it requires Optic neuritis, Acute myelitis, and at least two of three supportive criteria: Contiguous spinal cord MRI lesion extending over three vertebral segments; with Brain MRI meeting diagnostic criteria for MS

Because of the greater inflammatory nature in the pediatric age group as compared to adults all POMS patients should be started with Disease modifying Therapy (DMT). [8]

4. Conclusion

Acquired bilateral ptosis with ophthalmoplegia external should be given special attention to rule out systemic associations. Timely diagnosis and early management of multiple sclerosis in the paediatric age group with DMT may reduce relapse frequency and functional disability.

5. Conflict of Interest

None.

6. Source of Funding

None.

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