

A Rare Presentation of Isolated Oculomotor Nerve Palsy due to Multiple Sclerosis in a Child

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Abstract

Multiple sclerosis (MS) is an inflammatory, demyelinating, neurodegenerative disorder of the central nervous system (CNS) of unknown etiology. Isolated oculomotor nerve palsy is found rarely in children with MS. We present a case of MS in 10-year-old boy, who had bilateral ptosis associated with medial gaze restriction. The extraocular muscle examination demonstrated bilateral adduction palsy, impaired upward and downward deviation, and bilateral ptosis. Magnetic resonance imaging of brain revealed demyelinating changes in the midbrain.

Key Words: Child, Multiple Sclerosis, Oculomotor palsy, Third nerve.

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1- INTRODUCTION

Demyelinating disorders of the central nervous system (CNS) cause acute or relapsing-remitting encephalopathy and other multifocal signs of brain, brainstem, and spinal cord dysfunction (1). Demyelinating lesions within the brain stem often result in double vision and ocular motility disturbances, including nystagmus, Internuclear ophthalmoplegia (INO), and ocular motor cranial palsies. MS is being frequently diagnosed and it forms the differential diagnosis of many myelopathies (2, 3).

Furthermore, etiology and immunopathomechanic explanations of MS remain elusive. Here we report the rare clinical manifestations of multiple sclerosis such as isolated oculomotor nerve palsy in a child.

2- CASE REPORT

A 10- year –old- boy presented with complained of sudden onset drooping of eyelid. Three weeks before admission, ptosis of the left eyelid developed and during the next few days right eyelid ptosis developed as well. He had no headache, vomiting, convulsions, loss of consciousness, weakness or history suggestive of any other cranial nerve involvement.

He had no recent infectious or traumatic events and his personal and familiar medical history was otherwise unremarkable. On physical examination he was afebrile and hemodynamically stable. There was no evidence of lymphadenopathy, pallor, cyanosis or icterus. Examination of eye revealed ptosis, associated with medial gaze restriction (Left > Right). The result of

extraocular muscle examination demonstrated bilateral adduction palsy, impaired upward and downward deviation, and bilateral ptosis (**Figure.1**). Abduction was bilaterally intact. Bilateral papillary sizes were equal, as were bilateral responses to light. There was no nystagmus and other extraocular movements were normal. He had no other cranial nerve involvement or any other focal neurological deficit and there were no signs of meningeal irritation.

The remaining neurological examination, namely visual acuity, fundoscopic exam and long tracts systems were normal. Other system examination revealed no abnormality.

On investigations, his hemoglobin 10 g/dL with a total leucocyte count of 6,600 and a platelet count of 2.5lac/mm³. His blood sugars, renal and hepatic function tests were essentially normal. Chest X-ray revealed normal lung fields.

Erythrocyte sedimentation rate (ESR) was 25mm in 1st hr, Moux test was non-reactive. Cerebrospinal fluid examination revealed normal protein (28 mg/dL), and sugar (67mg/dL), with 3 lymphocytes, and no polymorphonuclear cells on cytology. Nerve conduction study was normal.

Magnetic resonance imaging of brain revealed area of altered signal intensity involving midbrain and pons appearing isointense on T1 weighted image (T1WI) and hyperintense on T2 weighted image (T2WI) suggestive of demyelination.

Oral steroid was given for 5days. His cranial nerve palsy right sided was slightly improved (**Figure.2**) before discharge.



Fig.1: Child with bilateral ptosis.



Fig.2: Child with slightly improved right sided cranial nerve palsy.

3- DISCUSSION

Pediatric MS is rare, with an estimated 2-5% of MS patients experiencing their 1st symptoms before age 18 years old. It has a female predominance by age 12 years. A complex interplay of environmental, infectious and genetic factors influence MS susceptibility (3). Isolated cranial nerve palsies are rare clinical signs in multiple sclerosis (2-5). Levy et al. (4)

have underlined that minor head trauma can cause an isolated oculomotor palsy in the absence of abnormal findings on brain MRI which was not the case of our patient. Presenting symptoms in pediatric MS include hemiparesis or paraparesis, unilateral or bilateral optic neuritis, focal sensory loss, ataxia, diplopia, dysarthria, or bowel/bladder dysfunction (3-6).

Patients with oculomotor nerve involvement may present with droopy lid, restricted ocular motility and binocular diplopia, if the ptosis is incomplete. The position of eye is down and out in isolated complete oculomotor nerve palsy same finding was present in our case. Associated symptoms are of extreme importance and patient should always be asked and assessed for any associated headache, periocular or orbital pain, visual involvement, red protruding eye, facial or body numbness and tremors, loss of smell or taste, involvement of trochlear and abducens nerves, loss of hearing, tinnitus or any other major ophthalmic or neurological involvement.

Ing et al. (7) found that trauma and bacterial meningitis accounted for more cases of isolated oculomotor nerve palsy than seen in the previous literature. Green et al. (8) reported that among 130 patients with isolated oculomotor nerve palsy only 2 patients had bilateral involvement; one had diabetes mellitus and the other had metastatic lung carcinoma.

Thomke et al. (9) found 24 patients with definite multiple sclerosis with isolated cranial nerve palsies during a 10 year period. Cranial nerve palsies were (third and fourth nerve: one patient each, sixth nerve: 12 patients, seventh nerve: 3 patients, eighth nerve: 7 patients).

The possibility of recurrent acute disseminated encephalomyelitis (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. MRI is the best method for showing dissemination in space, especially at the onset of multiple sclerosis, and is also thought to be the most sensitive method to detect demyelinating brainstem lesions (10-13).

Cranial MRI exhibits discrete T2 lesions in cerebral white matter, particularly periventricular regions as well as brainstem, cerebellum, and juxtacortical and deep gray matter. Cerebrospinal fluid (CSF), may be normal or exhibit mild pleocytosis, particularly in younger children. Abnormal MS profiles (increased IgG index, and/or CSF oligoclonal bands) increase likelihood of MS. CSF oligoclonal banding was not done in our cases, due to lack of such facility. Bhatti et al. (14) reported one MS patient with isolated III CN palsy caused by peripheral nervous system involvement that differently from our patient had pupillary involvement and enhancement of the cisternal portion of the oculomotor cranial nerve on MRI.

To the best of our knowledge not a single child was reported an isolated oculomotor cranial nerve paresis as the presenting sign of MS. Challenges arise in distinguishing pediatric MS from other demyelinating syndromes such as ADEM or neuromyelitis optica. 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 (3-5).

Relapses causing functional disability may be treated with methylprednisolone, 20-30 mg/kg/day) for 3-5 days, with or without prednisone taper. Lastly, 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. In conclusion, a high clinical awareness and neurological examination is necessary to the possibility of MS diagnosis.

4- CONCLUSION

Multiple sclerosis is an inflammatory, demyelinating, neurodegenerative disorder of the central nervous system of unknown etiology. Clinical awareness and neurological examination is necessary to

the suspect the possibility of MS diagnosis. A rare clinical manifestation of multiple sclerosis is isolated oculomotor nerve palsy.

5- CONFLICT OF INTEREST: None.

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