# An unusual case of isolated trochlear nerve palsy

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## ABSTRACT

Trochlear nerve palsy is rarely encountered in children and only 5% are truly isolated. Multiple sclerosis (MS) is also extremely uncommon in children. This report describes an otherwise healthy 10-year-old boy who presented with a 5-day history of vertical diplopia with associated dizziness, decreased appetite, and unsteadiness. He had no recent history of infection and no previous history of neurological symptoms. Ophthalmologic assessment revealed full ocular ductions and right hyperdeviation in primary gaze during alternate cover test. This hyperdeviation increased during left gaze, and right head tilt consistent with paresis of the right oblique muscle. Brain MRI revealed multiple well-defined hyperintense T2 lesions in the periventricular and subcortical white matter, and brainstem, suggestive of MS. His symptoms resolved after 2 weeks with no treatment. To conclude, isolated trochlear nerve palsy can be the initial clinical manifestation of childhood MS. Long term follow up is needed to confirm the diagnosis of MS.

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Fourth cranial (trochlear) nerve palsy results in weakness of the superior oblique muscle causing vertical diplopia that is invariably worse in downgaze.[1] Head injury is the principle cause, however, other causes include surgical injury such as anterior temporal lobectomy,[2] inflammatory process such as herpes zoster,[3] brain tumors such as schwannoma or metastases,[4,5] pseudotumor cerebri,[6] or vascular insult.[7] Trochlear nerve palsy is rarely encountered in children and only 5% of patients are truly isolated, namely, without other neurologic or ophthalmologic signs or symptoms.[1] Multiple sclerosis (MS) is also extremely uncommon in early childhood, particularly in those less than 10 years of age.[2] Although isolated cranial nerve palsies can be associated with MS, trochlear nerve palsy has been rarely reported.[3,4] To date only one child with documented trochlear nerve palsy, relapsing, and remitting neurologic deficits due to MS has been reported.[3] This report describes a child with isolated trochlear nerve palsy as an initial presentation of MS.

**Case Report.** An otherwise healthy 10-year-old boy was evaluated 5 days after he developed vertical diplopia. He had no recent history of upper respiratory tract infection or fever. He had associated dizziness, decreased appetite, and unsteadiness of gait. He had no previous history of weakness, sensory or vision loss. His medical history was unremarkable, and family history was negative for MS. Ophthalmologic assessment revealed full-appearing ocular ductions and right hyperdeviation in primary gaze during alternate cover test. This hyperdeviation increased during left gaze and right head tilt consistent with paresis of the right oblique muscle. The remainder of systemic and neurological examination was completely normal. Laboratory investigations revealed normal complete blood count, electrolytes, antinuclear antibody assay, liver, and renal function tests. Brain MRI revealed multiple well-defined asymmetrical hyperintense T2 lesions in the periventricular and subcortical white matter, splenium of the corpus callosum, and brainstem (**Figures 1-3**). No lesion was noted near the origin of the trochlear nerve. The lesions showed no contrast enhancement after intravenous gadolinium. An MRI of the orbits revealed normal optic nerves and ocular muscles. The parents refused lumbar puncture for CSF examination. His symptoms resolved completely after 2 weeks with no treatment. No further neurological signs or symptoms were encountered on follow-up.

**Discussion.** Isolated trochlear nerve palsy as an initial presentation of MS was not reported in pediatric patients. Jacobson et al,[3] described 5 patients (4 adults) with trochlear nerve palsy and MS. In 2 of these 5 patients, the trochlear nerve palsy was the initial clinical manifestation of MS. They reported on an 11-year-old boy who had multiple cranial

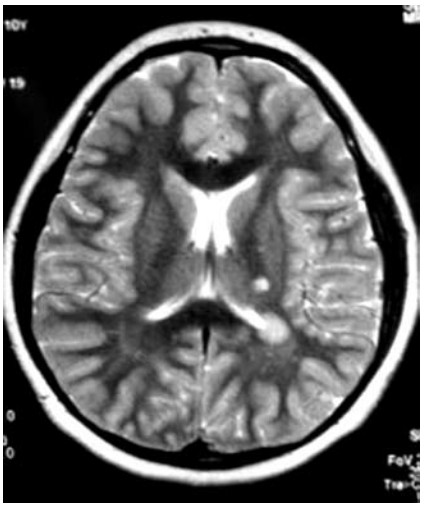

**Figure 1 -** Brain MRI showing multiple well-defined asymmetrical hyperintense T2 lesions in the periventricular and subcortical white matter.

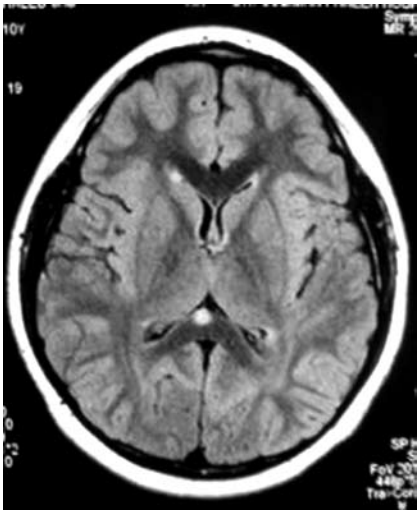

**Figure 2 -** The lesions also involved the corpus callosum.

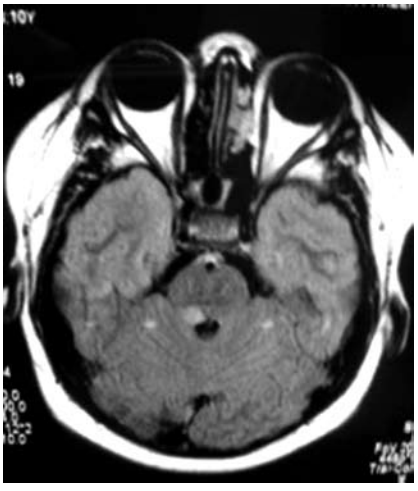

**Figure 3 -** Additional lesions involving the cerebellum and brainstem.

nerve involvements, namely, the trochlear nerve palsy was not isolated. In fact, other authors questioned the clinical diagnosis of trochlear nerve palsy in these reported cases.[5] Not every patient with vertical diplopia has trochlear nerve palsy. Other differential diagnoses that should be considered include third cranial nerve palsy, skew deviation, ocular myasthenia, and restrictive ophthalmoplegia in thyroid eye disease.[3,5] These disorders can be differentiated by history, ocular motility examination, and other associated findings. Skew deviation is a supranuclear vertical ocular misalignment due to disruption of the prenuclear input to the oculomotor and trochlear nerve nuclei. Skew deviation can be easily distinguished from the other causes of vertical diplopia because of the characteristic ocular misalignment that does not mimic other patterns of vertical ophthalmoplegia and because of the presence of other signs indicating brainstem dysfunction.[3]

The MRI findings in our patient are typical of MS; however, the child had no previous history of CNS signs or symptoms and remained normal on follow-up. Childhood MS is a demyelinating autoimmune disease with similar clinical and MRI findings to adult onset MS, but different genetic risk factors. Childhood MS is rare and adolescent presentation is most common.[6] Ghezzi et al,[7] found that of 149 children younger than 16 years presenting with MS, 73% presented with an initial attack after 13 years of age. No single laboratory finding is necessary or sufficient for diagnosing MS. Oligoclonal bands in the CSF were present in 75% of

**Table 1 -** Differentiating clinical, laboratory, and MRI features of ADEM and MS at initial presentation.[6,9,10]

| Feature | ADEM | MS |
|---|---|---|
| History of recent infection or vaccination | Common | Rare |
| ***Systemic symptoms*** | Common | Rare |
| Fever | 43% | 6% |
| Headache | 57% | 24% |
| Fatigue | 71% | 29% |
| Vomiting | 57% | 0% |
| Encephalopathy | 71% | 6% |
| Seizures | Common | Rare |
| Isolated optic neuritis | Less common (23%) | Common |
| Severity of illness | Usually severe | Less severe |
| CSF oligoclonal bands | 29% | 75% |
| ***MRI changes*** | | |
| Periventricular distribution | 50% | 91% |
| Involvement of the corpus callosum | 17% | 64% |
| Recurrence | Rare | Common |

ADEM - Acute disseminated encephalomyelitis, MS - Multiple sclerosis

children with clinically definite MS.[8] The parents of our child refused CSF examination. The diagnosis of MS in our patient remains probable. An alternate possibility is acute disseminated encephalomyelitis (ADEM). Several differentiating clinical, laboratory, and radiological features at initial presentation are summarized in **Table 1**.[6,9,10] Based on both clinical and MRI features, ADEM is less likely in our patient. Further attacks are required to confirm the diagnosis of MS, however, up to 14% of children with MS had the second attack more than 10 years after the initial presentation.[6] To conclude, isolated trochlear nerve palsy can be the initial clinical manifestation of childhood MS. Long term follow up is needed to confirm a clinical diagnosis of MS.

## References


1. Keane JR. Fourth nerve palsy: historical review and study of 215 inpatients. ***Neurology*** 1993; 43: 2439-2443.
2. Tardieu M, Mikaeloff Y. Multiple sclerosis in children. ***Int MS J*** 2004; 11: 36-42.
3. Jacobson DM, Warner JJ, Ruggles KH. Transient trochlear nerve palsy following anterior temporal lobectomy for epilepsy. ***Neurology*** 1995; 45: 1465-1468.
4. Thomke F, Lensch E, Ringel K, Hopf HC. Isolated cranial nerve palsies in multiple sclerosis. ***J Neurol Neurosurg Psychiatry*** 1997; 63: 682-685.
5. Lavin PJ, Donahue SP. Isolated trochlear nerve palsy in patients with multiple sclerosis. ***Neurology*** 2000; 55: 321-322.
6. Jones CT. Childhood autoimmune neurologic diseases of the central nervous system. ***Neurol Clin N Am*** 2003; 21: 745-764.
7. Ghezzi A, Deplano V, Faroni J. Multiple sclerosis in childhood: clinical features of 149 cases. ***Mult Scler*** 1997; 3: 43-46.
8. Selcen D, Anlar B, Renda Y. Multiple sclerosis in childhood: report of 16 cases. ***Eur Neurol*** 1996; 36: 79-84.
9. Brass SD, Caramanos Z, Santos C, Dilenge ME. Multiple sclerosis vs acute disseminated encephalomyelitis in childhood. ***Pediatr Neurol*** 2003; 29: 227-231.
10. Mikaeloff Y, Suissa S, Vallee L, Lubetzki C. First episode of acute CNS inflammatory demyelination in childhood. Prognostic factors for multiple sclerosis and disability. ***J Pediatr*** 2004; 144: 246-252.


## *CASE REPORTS*

Case reports will only be considered for unusual topics that add something new to the literature. All Case Reports should include at least one figure. Written informed consent for publication must accompany any photograph in which the subject can be identified. Figures should be submitted with a 300 dpi resolution when submitting electronically or printed on high-contrast glossy paper when submitting print copies. The abstract should be unstructured, and the introductory section should always include the objective and reason why the author is presenting this particular case. References should be up to date, preferably not exceeding 15.