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# A Case of Low-Grade Astrocytoma First Thought to be Demyelinating Disease on the Basis of Initial Cranial MR Findings

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### Abstract

Oligodendrogliomas are rare childhood brain tumors. Cranial MR images are nonspecific. Identification is easier with histopathological and genetic investigation. We report a 15-month-old male patient presenting with restlessness and inability to walk over the preceding week. Although cranial MR findings suggested demyelinating white matter disease, oligodendroglioma was diagnosed on the basis of biopsy following radiological findings. We present this case in order to describe the initial radiological findings and to emphasize the importance of MR spectroscopy in diagnosis in such patients.

**Keywords:** MR spectroscopy; Oligodendroglioma; Demyelinating white matter disease; Schilder's disease

# Introduction

Clinical findings at presentation in childhood gliomas may be confused with demyelinating diseases, and particularly with ADEM. Clinical findings at presentation may be similar in both. In addition to symptoms such as sudden onset seizure, headache and vomiting, patients may also present with other neurological complaints, such as gradual weakness and gait impairment [1-3]. MR images make a significant contribution to diagnosis in both disease groups. The possibility of early diagnosis in glial tumors has increased with the use of MRI and stereotactic biopsy. We report a 15-month-old male patient presenting to our clinic with gait impairment. Clinical findings and initial cranial MR images suggested demyelinating disease, and steroid therapy was administered. Symptoms improved following steroid therapy. Oligodendroglioma was diagnosed on the basis of control cranial MR, MR spectroscopy and biopsy.

### **Case Report**

A 15-month-old male patient presented to our clinic with gait impairment and restlessness. Trembling in the hands and feet when the patient was restless had troubled the family for some time. Although he had begun walking normally by holding onto objects, in the preceding few days he had only been able to remain standing for 1-2 seconds. No findings other than restlessness were present at physical examination. Blood biochemistry and acute phase reactants were normal. Craniospinal MR was performed. Hyperintense lesions in the right temporoparietal and left parietal regions were interpreted as inflammatory demyelinating pathology (Figure 1). No cells were



**Figure 1:** Transverse unenhanced FLAIR MR images (a and b) show hyperintense lesions in right temporoparietal and left parietal lobe. There is no mass effect and apparent peripheral edema.



**Figure 2:** Follow-up images, performed by 5 months later, axial T2-weighted MR image (a) and transverse unenhanced FLAIR MR image (b) reveal progression of right temporoparietal hyperintense lesion.

observed in CSF. CSF protein was 23 mg/dl. Restlessness decreased following pulse steroid therapy, and the patient began walking. CSF NMDA receptor antibody Anti GAD was negative. Mass lesion in the central nervous system was suspected at repeat cranial MR (Figure 2). No contrast involvement was observed (Figure 3). MR spectroscopy showed elevated choline (Cho) and decreased N-acetyl aspartate (NAA) (Figure 4). Biopsy results were interpreted as low-grade glioma. Chemotherapy was administered following surgery. The patient recovered after chemotherapy. There was no recurrence of the problem with the patient's gait. Pathological findings and genetic studies when the residual mass was totally excised approximately 10 months later were compatible with grade 2 oligodendroglioma (Figure 5a and 5b).

# Discussion

Brain tumors constitute the second largest group of childhood

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Page 2 of 3

cancers after leukemias in children aged 0 to 14. Leukemias comprise 30% of cases, while tumors of the central nervous system occur in 20% to 24% of patients [4]. Oligodendrogliomas constitute 5% to 18% of childhood gliomas and 1% of childhood brain tumors [5,6].



Figure 3: Initial contrast-enhanced axial T1-weighted MR image does not show contrast enhancement within the lesion.



Figure 4: MR spectroscopy image shows elevated choline (Cho) and decreased N-acetyl aspartate (NAA). This finding is consistent with neoplasm.



**Figure 5a:** (HE X 20) Places monitored thin capillary generally oval-round core fine chromatin in cells showing mild atypia within fibrillary stroma containing vessels. Noteworthy structures were microcystic.



Figure 5b: (HE X40) Oval-spherical fine chromatin core cells is observed perinuclear clear halo around some cell nuclei.

The epidemiological prevalence of pediatric white matter disorders as a class are not well defined. Disorders tend to be classified as acquired demyelinating disorders, leukodystrophies or heritable leukoencephalopathies. Acquired demyelinating disorders include multiple sclerosis, neuromyelitis optica, transverse myelitis, optic neuritis and acute disseminated encephalomyelitis. Schilder's disease is a rare variant of multiple sclerosis generally seen in children. Demyelinating diseases, and particularly Schilder's disease, were considered at differential diagnosis in our case based on the initial cranial imaging. Schilder's disease is a rare disorder characterized by an inflammatory white matter plaque of demyelination. At MR of the brain, lesions are hypodense on T1 sequences and hyperintense on T2 [7]. Although oligodendrogliomas may be seen in all regions of the brain, they generally involve white matter in the cerebral hemispheres [8-11]. Hypointense signal changes are observed on T1 sequences and hyperintense changes on T2/FLAIR. Post-contrast enhancement may also be seen [12]. MRS findings are nonspecific, showing elevated choline and low NAA levels, as with many other tumors. Clinical improvement after high-dose steroid therapy supported our initial diagnosis for some time. Tumors of the central nervous system can be confused with clinical findings of childhood demyelinating diseases. This is because demyelinating diseases, and particularly ADEM, are widely seen in this age group [13]. Presence of viral infection and a history of vaccination before these diseases and sudden and severe onset of clinical symptoms can assist differentiation from tumors of the central nervous system. Although MR findings generally make a major contribution to diagnosis, there may be conditions, as in this case, in which differentiation is problematic. CSF findings may sometimes assist diagnosis. As in this case, high-dose steroid therapy administered to treat demyelinating diseases may sometimes delay diagnosis by causing a temporary decrease and improvement in clinical symptoms due to the effect of the mass. Tumors of the central nervous system should be considered at differential diagnosis under such circumstances. Tumors of the central nervous system may sometimes mimic childhood demyelinating diseases. In addition to close clinical and neuroradiological monitoring, early MR spectroscopy is also required in such circumstances.

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Page 3 of 3