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Tumoral Multiple Sclerosis of the Cerebellum in a Child

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Summary: We present a case of cerebellar tumoral multiple sclerosis in an 11-year-old girl and emphasize these two features: (1) Tumoral multiple sclerosis can occur within the posterior fossa and should be strongly considered in the differential diagnosis of mass lesions of the posterior fossa when typical white matter lesions are seen on T2-weighted images. (2) Tumoral multiple sclerosis can demonstrate ring enhancement on MR.

Index terms: Sclerosis, multiple; Cerebellum, neoplasms; Pediatric neuroradiology

Tumoral multiple sclerosis is an uncommon manifestation of multiple sclerosis and is rarely reported in children (1). Documented cases to date have occurred within the cerebrum, even though multiple sclerosis plaques are commonly found within the cerebellum and brain stem (2). As a result, tumoral multiple sclerosis has been an unlikely consideration in the differential diagnosis of cerebellar mass lesions in any age group, although, as the case we report demonstrates, a mass of the posterior fossa associated with multiple white matter lesions on T2-weighted magnetic resonance (MR) images should make one strongly consider the diagnosis of tumoral multiple sclerosis. We report an 11-year-old girl who had right cerebellar tumoral multiple sclerosis.

Case Report

An 11-year-old right-handed girl was admitted to our institution because of a right cerebellar mass. She had a 2-week history of intermittent headache, nausea, and vomiting and a 1-week history of head tilt, poor balance, and deteriorating handwriting ability. She had no recent vaccinations or viral illnesses. When she was 4 years old, an episode characterized by ataxia, hand tremor, falling to the right, and difficulty walking was thought to be caused by viral encephalitis. A computed tomographic scan of the

brain at that time was reported to be normal. She made a full recovery after that event.

On physical examination she had right lateral nystagmus, right dysmetria with dysidiadochokinesia, and incoordination of right lower extremity movement. Ophthalmologic examination was normal. MR imaging of the brain showed a right cerebellar hemispheric mass involving the middle cerebellar peduncle and extending into the hemispons. Moderate mass effect was present causing deformity of the fourth ventricle but not hydrocephalus. The lesion had low signal intensity on T1-weighted images and high signal intensity on T2-weighted images (Fig 1A). Extensive edema surrounded the lesion. After injection of gadopentetate dimeglumine, an irregular enhancing ring (Fig 1B) was present. Other lesions were observed: in the left cerebellar hemisphere, a bright lesion near the cerebellopontine angle was seen on T2-weighted images (Fig 1A); in the cerebrum, several bright ovoid periventricular lesions (Fig 1C) were seen on T2-weighted images. Two of the latter showed enhancement on post contrast T1-weighted images. Because of the multiplicity of lesions and the rapid clinical evolution of the mass, the possibility of a high-grade malignant neoplasm was initially considered. However, findings on thallium single-photon emission computed tomography were normal.

Biopsy and debulking of the right cerebellar mass was planned. However, frozen sections revealed abnormal cerebellar tissue indistinguishable from low-grade glioma. Therefore, subtotal resection of the mass was performed. Histopathologic examination of the final surgical specimen showed an extensive demyelinating process of the white matter associated with diffuse perivascular infiltration by lipid-containing macrophages. Focally, perivascular lymphocytic infiltrates were present. Luxol fast blue periodic acid-Schiff stain depicted loss of myelin in the major portion of the lesion with scant amounts of myelin at the transition to normally myelinated white matter. The findings were also confirmed by electron microscopy. The final diagnosis was a demyelinating process consistent with multiple sclerosis.

Further testing including lumbar puncture was refused by the parents. Repeat brain MR imaging 7 months later showed decreased mass effect of the cerebellar lesion as

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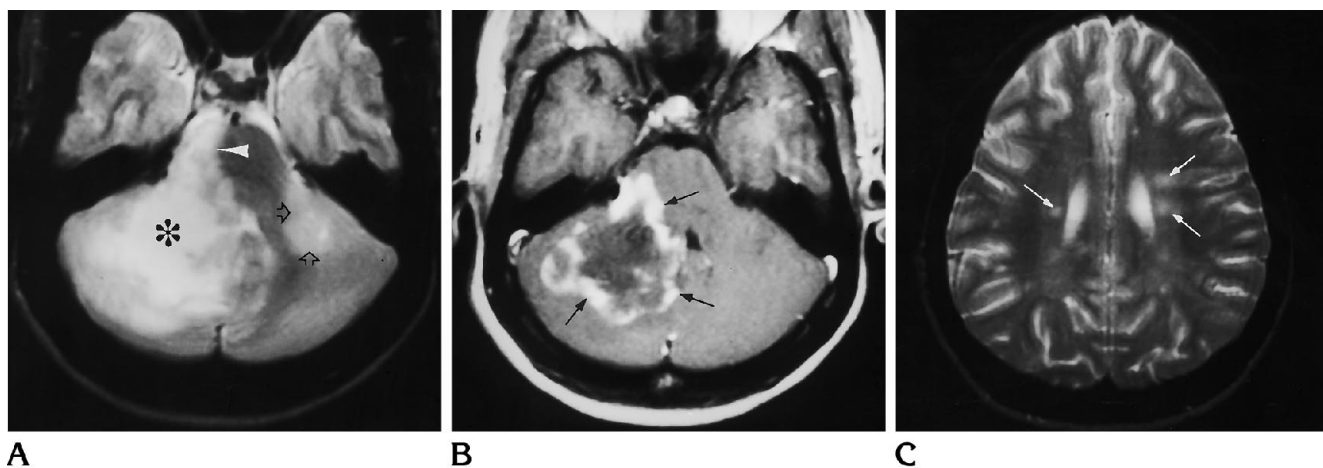


Fig 1. 11-year-old girl with cerebellar tumoral multiple sclerosis.

A, Axial T2-weighted MR image (2320/90/2 [repetition time/echo time/excitations]) shows increased signal within right cerebellar tumoral multiple sclerosis lesion (*asterisk*) with surrounding edema, mass effect on the fourth ventricle, and extension into the right hemispheres (*arrowhead*). Left cerebellar multiple sclerosis plaque is also shown (*arrows*).

B, Axial T1-weighted MR image (584/18/1) after contrast administration shows irregular ring enhancement of the right cerebellar lesion (*arrows*).

C, Axial T2-weighted MR image (2390/90/20) shows several periventricular multiple sclerosis plaques oriented perpendicular to the lateral ventricles (*arrows*).

well as two new bright lesions on T2-weighted images in the right centrum semiovale. Clinically, the patient was doing well with no recurrence of symptoms.

Discussion

Multiple sclerosis is a common demyelinating disease most frequently encountered in young and middle-aged adults. Clinical manifestations are variable but commonly include visual, motor and/or sensory changes, and cerebellar dysfunction (3). The clinical course is unpredictable, but most often there is an incremental progression with exacerbations and remissions (2). Diagnosis is commonly based on the criteria of Poser et al (4).

Characteristic MR findings consist of multiple white matter lesions with low signal on T1-weighted images and bright signal on T2-weighted images (2, 5). These lesions are observed most often in the periventricular white matter and the corona radiata, where they can be ovoid with their long axes perpendicular to the ventricular walls (6). Other common locations include the centrum semiovale, corpus callosum, brain stem, corpus medullaris, and middle cerebellar peduncles (2). Active lesions frequently enhance with contrast material (2, 5, 7, 8) and have displayed ring enhancement on computed tomography, with the ring thought to represent the active edge of demyelination (7).

Multiple sclerosis in children and adolescents demonstrates several remarkable differences when compared with adults. First, the initial childhood presentation frequently simulates acute disseminated encephalomyelitis, cerebral abscess, or an acute metabolic encephalopathy, presentations rarely seen in adults (9). Second, children with multiple sclerosis have a 10% mortality during the first 5 years of the disease, compared with adults, who have 0% mortality over a similar time period (9). Third, there is a 5:1 female predominance of multiple sclerosis in the adolescent population, compared with 1.4–1.9:1 in adults (10). Fourth, adolescents have cerebellar and brain-stem plaques much more often (70% and 80%, respectively) than adults do (33% and 14%, respectively) (10). Finally, abnormal iron deposition within the basal ganglia, as well as cortical atrophy, is uncommon in adolescents with multiple sclerosis but is common in adults (10).

The tumoral form is an uncommon presentation of multiple sclerosis that is believed to represent a fulminant acute demyelinating plaque or conglomeration of acute plaques forming a mass lesion (2). On noncontrast computed tomography, it is usually hypodense. After the administration of iodinated contrast material, it may display ring enhancement simulating tumor or abscess (11). With MR, tumoral multiple

sclerosis usually displays low signal on T1-weighted images and bright signal on T2-weighted images (11, 12) when compared with normal white matter. Mild surrounding edema and mass effect are typical (12). When such a lesion is found in association with typical white matter lesions, the diagnosis of tumoral multiple sclerosis should be strongly considered. The differential diagnosis of solitary tumoral multiple sclerosis includes tumor, abscess, and possibly a tumoral demyelinating entity separate from multiple sclerosis and acute disseminated encephalomyelitis that was described by Kepes (13). Tumoral multiple sclerosis associated with bright periventricular lesions on T2-weighted images needs to be differentiated from disseminated glioma or gliomatosis cerebri and the rare association of multiple sclerosis with coincident glioma (14). Because cerebellar astrocytomas are common tumors of childhood, the occurrence of posterior fossa tumoral multiple sclerosis takes on special significance.

There have been few documented cases of tumoral multiple sclerosis in children (1, 12). Radiographically, there has been no difference between pediatric and adult cases. Our case is distinctly atypical because it arose from the right cerebellar hemisphere and adjacent middle cerebellar peduncle. The clinical history of multiple episodes with recovery, the appearance and enhancement characteristics of the periventricular lesions, and the development of new lesions on follow-up confirm the diagnosis of multiple sclerosis and negate the diagnosis of acute disseminated encephalomyelitis. The tumoral demyelinating entity described by Kepes (13) is unlikely because his criteria included improvement on follow-up imaging studies without development of new lesions.

Nesbit et al (5) presented enhanced MR findings of a large frontal lobe lesion of multiple sclerosis accompanied by significant mass effect and homogeneous enhancement. The irregular MR ring enhancement in our case is different from the case of Nesbit et al but similar to the ring enhancement on computed tomography described by Gutling and Landis (11). Our pathologic findings, demonstrating active demyelination at the transition zone of the lesion, tend to support the hypothesis of Miller et al (7) that the enhancing ring corresponds to the active demyelination seen at the edge of the plaque.

In summary, we present the case of an 11-year-old girl who had tumoral multiple sclerosis arising from the cerebellum, an unusual location for this entity. In addition, this case confirms the occurrence of MR ring enhancement in tumoral multiple sclerosis. The differential diagnosis for a child or adolescent with a cerebellar mass should be expanded to include tumoral multiple sclerosis, especially in children and adolescents who have a history and other MR findings, such as multiple bright periventricular lesions on T2-weighted images, suggestive of multiple sclerosis.

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