Multiple Cranial Nerve Enhancement: A New MR Imaging Finding in Metachromatic Leukodystrophy

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Metachromatic leukodystrophy (MLD) is a set of several disorders caused by deficient lysosomal activity. This deficiency results in accumulation of a metachromatic lipid material, galactosylsphingosine sulfatide, leading to the breakdown of the myelin sheath in both central and peripheral nervous systems, initially sparing the subcortical “U” fibers. Among leukodystrophies, cranial nerve involvement has been documented in infantile Krabbe disease. Proposed mechanisms for the abnormal nerve root enhancement have included alteration of vascular permeability with breakdown of the blood-nerve barrier as a result of perivascular inflammation or infiltration and enhancement in areas of active myelin breakdown.

We presume that multiple cranial nerve enhancement in MLD is secondary to the accumulation of the metachromatic lipid material and to the disruption of the myelin sheath, similar to the process that affects peripheral nerves. A future investigation with a larger series is required to evaluate the usefulness of cranial nerve MR imaging analysis in suspected MLD patients.

References

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Congenital Glioependymal Cyst Presenting with Severe Proptosis

We discuss a unique case of a congenital glioependymal cyst arising from the left middle cranial fossa with extension into the left orbit causing severe proptosis, a presentation that has not been reported previously. A female neonate at 38 weeks’ gestational age was diagnosed with severe left proptosis on a prenatal sonography at 35 weeks’ gestation. The sonogram was performed to determine the size of the infant due to a maternal history of macrosomia in a previous pregnancy. The infant’s prenatal history was otherwise unremarkable. The patient was delivered by an uncomplicated cesarean birth.

Physical examination showed severe left proptosis with intact extracranial movements. The right eye and findings from the remaining cranial nerves were normal. Noncontrast enhanced CT demonstrated a cystic structure occupying the left middle cranial fossa and extending into the left orbit. MR imaging verified the CT findings and showed no associated brain abnormalities (Fig 1).

On the patient’s second day of life, a cyst-to-peritoneum shunt was placed without complications. The left proptosis rapidly improved, and a sonogram showed only a small cyst on the fifth day of life. The patient was discharged but was returned on the 15th day of life because of recurrent left proptosis. After a failed shunt revision, a left temporal craniotomy was performed with removal of the lateral wall of the cyst and fenestration of the cyst to the subarachnoid space.

Fig 1. A, Axial T2-weighted image shows the white matter involvement with a “tigroid” pattern. B, Coronal postcontrast T1-weighted image shows bilateral and symmetrical abnormal Gd enhancement of the oculomotor (arrowheads) and trigeminal nerves (arrow).