

UNILATERAL CEREBELLAR HYPOPLASIA- INCIDENTAL FINDING IN TWO CASES

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ABSTRACT

Unilateral cerebellar hypoplasia is a relatively rare malformation. We report two cases of unilateral cerebellar hypoplasia that were detected following fever and fits in a previously asymptomatic child.

Case Reports

Case 01: A one -year-old male child presented with a history fever for last four days associated with one episode of fits. MRI brain was advised for work up of fits. His birth history was insignificant with no history of birth asphyxia. Developmental history was also normal.

On examination, no neurologically significant complains. Cerebellar signs were not present.

An MRI scan showed hypoplasia of the left cerebellar hemisphere, a large cisterna magna, and asymmetry of the posterior fossa: the left side being smaller than the right (Fig. 1) some asymmetry of midbrain was also noted. Rest of brain imaging was normal. Based on these imaging findings, a diagnosis of unilateral cerebellar hypoplasia was made. The child was managed conservatively for fever and fits and he recovered.

Case 02: Another case of one and half year old boy presented with a history fever for last three days associated with one episode of fits. MRI brain was advised for work up of fits.

His birth history was insignificant, no history of birth asphyxia. Developmental history was also normal. On examination, no neurologically significant complains.

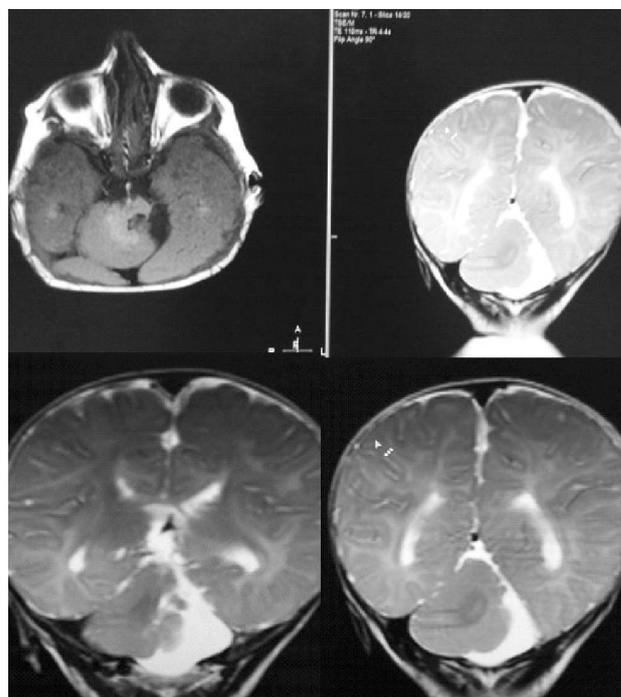


Figure 1: T1W coronal MRI images show left cerebellar hypoplasia (arrows).

Cerebellar signs were not present.

An MRI scan showed hypoplasia of the right cerebellar hemisphere and asymmetry of the posterior fossa: the left side being smaller than the right (Fig. 2). Rest of brain imaging was unremarkable. Based on

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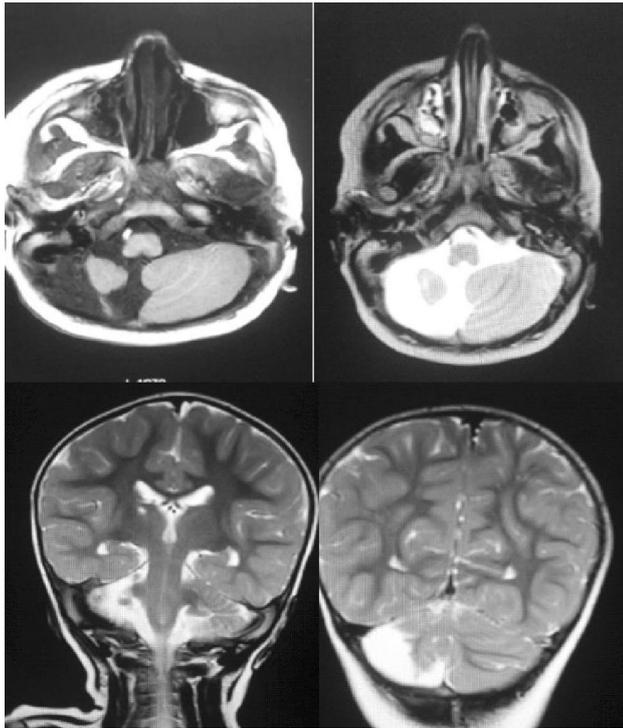


Figure 2: T1W coronal MRI images show right cerebellar hypoplasia

these imaging findings, a diagnosis of unilateral cerebellar hypoplasia was made. The child was managed conservatively for fever and fits and he recovered.

Discussion

On the basis of MRI findings, cerebellar malformations can be divided into those associated with hypoplasia and those with dysplasia; each type can show either focal or diffuse malformations. Focal cerebellar hypoplasia can be further subdivided into isolated vermis hypoplasia or hypoplasia of one cerebellar hemisphere.^{1,3,4} Pathologic evidence of cerebellar injury due to birth asphyxia is well described and, because of its high metabolic activity, the vermis is the structure that is most commonly involved.⁵ The clear demonstration of cerebellar hypoplasia, associated with hypoplasia or aplasia of the cerebellar or vertebral arteries, favors the concept of an intrauterine vascular etiology for cerebellar hypoplasia / aplasia.^{2,6} Genetic mutations with somatic mosaicism may also have a role to play.⁷ Children with cerebellar hypoplasia may

present with developmental delay especially gross motor delay and hypotonis.⁸ However, unilateral cerebellar hypoplasia may be an incidental finding in a patient with no previous evidence of neuromuscular or metabolic disease and no past history of trauma or anoxia. This was especially true in both of our cases.⁹

Long term outcome in different cases is variable ranging from no neurological complain to ataxia and developmental delay. It seems that involvement of the cerebellar vermis is associated with a poorer cognitive outcome, whereas an intact vermis is associated with normal cognitive outcome and no truncal ataxia.¹⁰

CT scan shows posterior fossa asymmetry with underlying unilateral cerebellar hemisphere hypoplasia. When available, MRI angiography may demonstrate the vascular anomalies in the cerebellar and / or vertebral arteries in most of the patients. In the present cases, the child's symptoms were primarily due to the fever, which responded well to conservative management. however as both these cases presented with fits and fits are associated with cerebellar hypoplasia according to literature, these patients are on regular follow up of Neurology department of Children Hospital. Follow up reveals that up till now no further fits occur in both these cases. From this we may conclude that cerebellar hypoplasia was an incidental finding in both of our cases.

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