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Clinical-Medical Image

Rhombencephalosynapsis: An Uncommon Cerebellar Malformation

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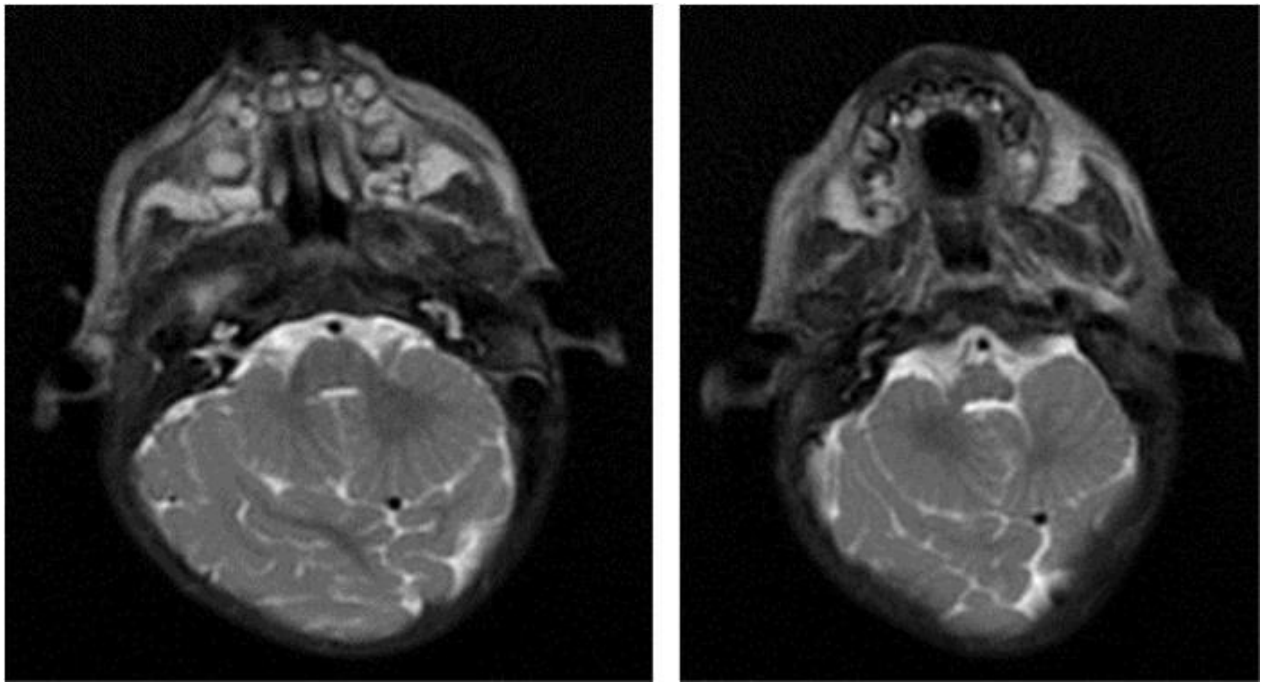

Figure 1: Cerebral MRI on axial T2 weighted-images showing complete vermian agenesis.

Clinical Image

We present the case of a 10 months old girl, who presented with a myelomeningocele. Brain MRI was performed along and showed complete vermian agenesis consistent with rhombencephalosynapsis associated with ventriculomegaly and corpus callosum hypoplasia. Rhombencephalosynapsis is a rare congenital malformation of the posterior cranial fossa, characterized by partial or total vermian agenesis, dorsal fusion of the cerebellar hemispheres and dentate nuclei, variably associated with fusion of colliculi and superior cerebellar peduncles [1]. Oberstein er first described this malformation in 1914, based on a postmortem examination of a 28 years old man [2].

The exact cause of this sporadic anomaly is still unknown. Some authors suggest that it results from a failure of vermian differentiation occurring between the 28^{th} and 44^{th} day of gestation [3].

This condition is commonly associated with other midline malformations such as ventriculomegaly, commissural hypoplasia of the commissural system, absence of the olfactory tract agenesis of the posterior lobe of the pituitary and hypoplasia of the anterior visual pathway [4,5]. Association with VACTREL and Gomez-Lopez-Hernandez syndromes has also been reported [6]. Clinical presentation is extremely variable as it depends on the associated supratentorial anomalies [6]. Prenatal diagnosis of this anomaly is currently possible. The presence of ventriculomegaly with cerebellar hypoplasia should prompt careful evaluation for rhombencephalosynapsis [7].

Magnetic resonance imaging (MRI) is the modality of choice to explore the posterior fossa. Typical findings include transversely oriented cerebellar folia and fissures across the midline without the intervening vermis, together with a diamond-shaped fourth ventricle. Thanks to its high spatial and contrast resolution, MRI is also able to assess the midline abnormalities often associated with this malformation [8].

Keywords: Rhombencephalosynapsis; Cerebellum; Vermis; Infant

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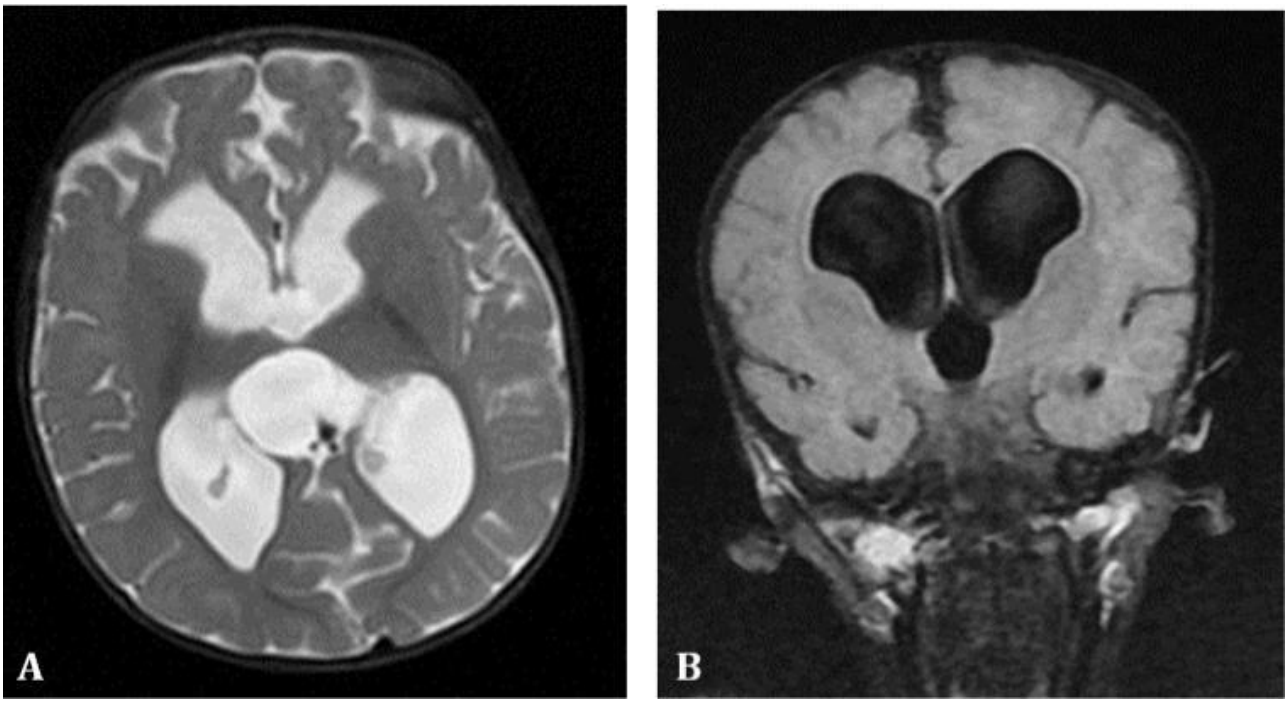

Figure 2: Cerebral MRI on axial T2 (A) and FLAIR (B) weighted-images showing ventriculomegaly and corpus callosum hypoplasia

Declaration of Interests

The authors declare that they have no competing interests.

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