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DANDY–WALKER MALFORMATION PRESENTING WITH MACROCEPHALY IN A TERM NEONATE: A CASE REPORT

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ABSTRACT

Dandy–Walker malformation (DWM) is a rare congenital brain malformation characterized by cystic dilatation of the fourth ventricle, hypoplasia or agenesis of the cerebellar vermis, and enlargement of the posterior fossa. We report a day-2 term male neonate presenting with macrocephaly and antenatal suspicion of DWM. Neuroimaging confirmed the diagnosis. The neonate remained clinically stable during NICU stay. Early diagnosis and multidisciplinary follow-up are essential to optimize long-term outcomes.

INTRODUCTION

Dandy–Walker malformation (DWM) is a rare congenital anomaly involving the cerebellum and fourth ventricle. It is characterized by cystic dilatation of the fourth ventricle, hypoplasia or agenesis of the cerebellar vermis, and enlargement of the posterior fossa. It is often associated with hydrocephalus and systemic anomalies. Early diagnosis using neuroimaging is essential for monitoring and timely intervention.

CASE REPORT

A day-2 term male neonate with birth weight of 3.5 kg (appropriate for gestational age) was delivered by LSCS with antenatal suspicion of Dandy–Walker malformation. The baby cried immediately after birth and was referred to a tertiary care NICU for further evaluation.

On examination, the neonate had macrocephaly with a wide anterior fontanelle measuring 6.5 × 7 cm. Systemic and neurological examinations were otherwise normal with normal tone and activity.

Neurosonogram revealed an enlarged posterior fossa with vermian hypoplasia and mild prominence of the right ventricle. MRI brain confirmed a large cystic lesion in the posterior fossa communicating with the fourth ventricle, consistent with Dandy–Walker malformation.

Screening echocardiography revealed a small ostium secundum ASD and PDA. The neonate remained hemodynamically stable without signs of raised intracranial pressure or respiratory distress during the NICU stay.

DISCUSSION

Dandy–Walker malformation is a congenital anomaly with variable clinical presentation. Macrocephaly is often an early clinical sign. Neuroimaging plays a crucial role in confirming the diagnosis and identifying associated anomalies.

Early identification allows for close monitoring of hydrocephalus and neurodevelopmental outcomes. Associated cardiac anomalies, as seen in this case, further highlight the importance of systemic evaluation.

Management is primarily supportive, with surgical intervention reserved for cases with progressive hydrocephalus.

CONCLUSION

Early neuroimaging is essential in neonates presenting with macrocephaly and suspected posterior fossa anomalies. Careful monitoring for hydrocephalus and developmental delay is necessary. Multidisciplinary follow-up involving neurosurgery, ophthalmology, and developmental specialists is crucial for improving long-term outcomes.

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