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Hemorrhagic Occipital Meningocele in Dandy Walker Syndrome: A Rare Case

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ABSTRACT

Dandy walker syndrome is commonest congenital cerebellar malformation with estimated prevalence of 1:25,000 to 1:50.000 live births. However, its association with occipital meningocele is a rarely seen. Here we present an unusual case of preterm neonate with Dandy walker syndrome and occipital meningocele, intraventricular haemorrhage and haemorrhage within the posterior fossa cyst. The patient was diagnosed antenatally in the third trimester and had a vaginal delivery.

Keywords: Dandy walker syndrome, Occipital meningocele, Intracranial haemorrhage

CASE REPORT

A 30-year-old G3 P1L1A1 patient, presented for first sonological evaluation at 35 weeks period of gestation. USG revealed vermian hypoplasia with a large posterior fossa cyst and occipital encephalocele. The cyst was seen communicating with dilated 4th ventricle across a defect in the roof of the 4th ventricle. Club foot deformity was also noted on the left side. There was overriding of the 2nd toe on the right side. Patient had a preterm labour with vaginal delivery at 35 weeks 5 days POG.

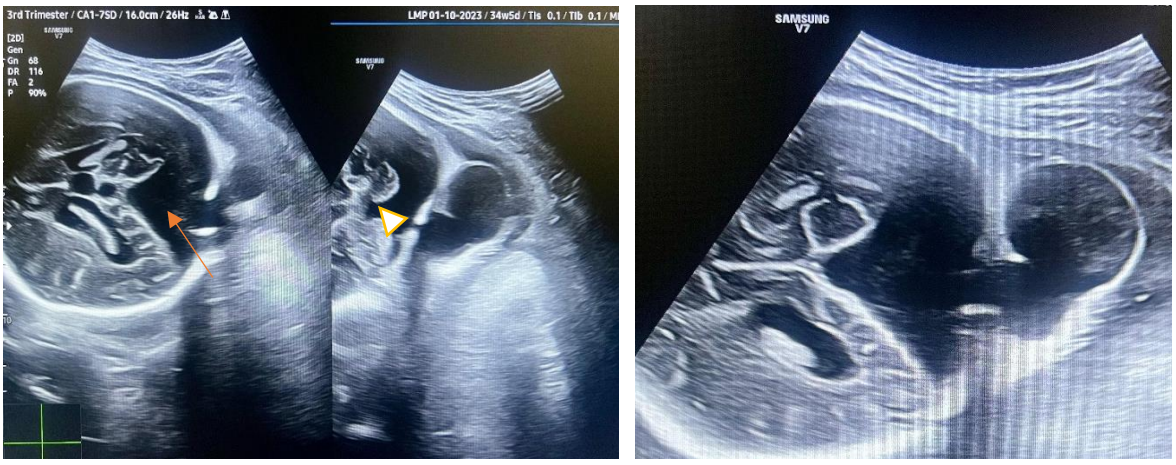

Figure 1: Antenatal ultrasound showing a large posterior fossa cyst (arrow), a defect in roof of the 4th ventricle (arrow head) and occipital calvarial defect with meningocele (right image).

On clinical examination, an osseous defect was noted in the occipital region with protrusion of a soft swelling overlying the defect. A caput succedaneum was noted. The

neonate also had birth trauma fracture clavicle.

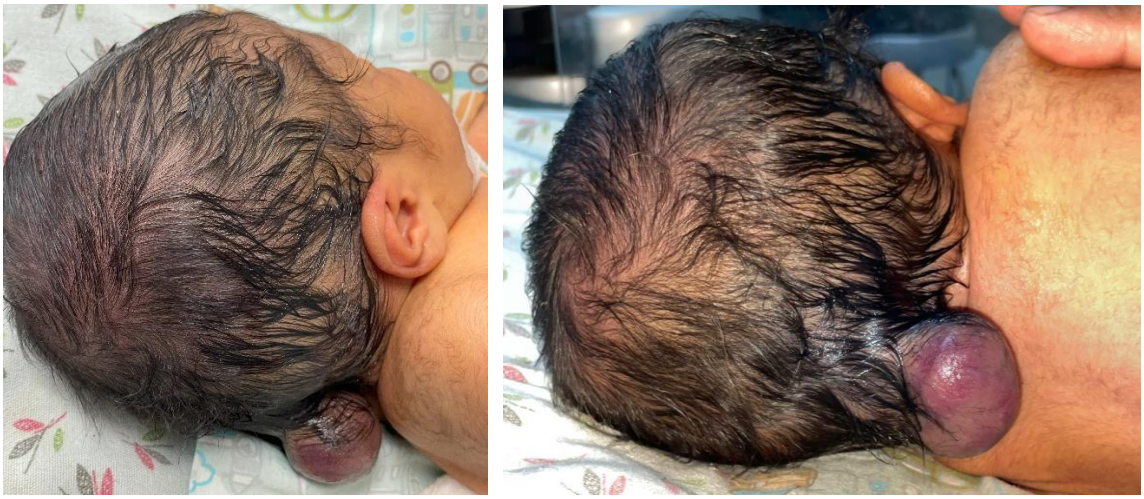

Figure 2: A soft violaceous swelling in the occipital region of the neonate covered with thinned out skin.

On initial imaging by transcranial sonography across the calvarial defect in the occipital region, a large posterior fossa cystic lesion which was seen communicating with the 4th ventricle. The cystic lesion was protruding out of the occipital calvarial defect. Echogenic floaters were noted in this large cystic lesion. There was intraventricular haemorrhage with no obvious dilatation of the ventricles.

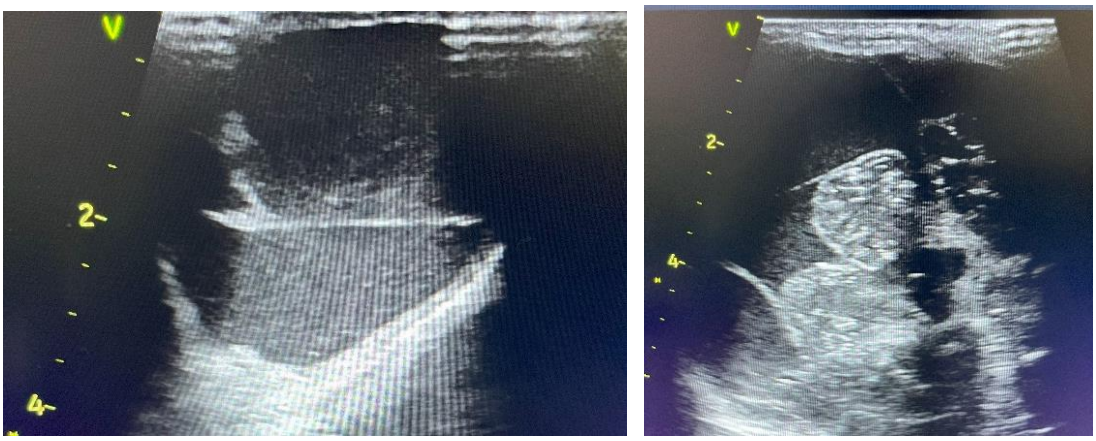

Figure 3: On ultrasound examination the occipital swelling appeared cystic with a few thin membranes and echogenic floaters with fluid-fluid level (Left). A large posterior fossa cyst with echogenic floaters was seen across the mastoid fontanelle (Right).

On MRI brain with screening of whole spine, there was a large cystic lesion in the posterior cranial fossa communicating anteriorly with the 4th ventricle across a defect in the inferior medullary velum. The cystic lesion was protruding posteriorly across a calvarial defect in the occipital region. It had blood CSF level, as evidenced by dependent area appearing hypointense on T2 weighted images, hyperintense on T1 weighted images with magnetic susceptibility artefacts on SWI images. A focal area of T1 shortening appearing hypointense on T2WI and blooming noted in the right centrum semiovale-consistent with ICH. Dependent areas of T1 hyperintensity, T2 hypointensity with blooming noted in the occipital horns of bilateral lateral ventricles-suggestive of intraventricular extension of bleed.

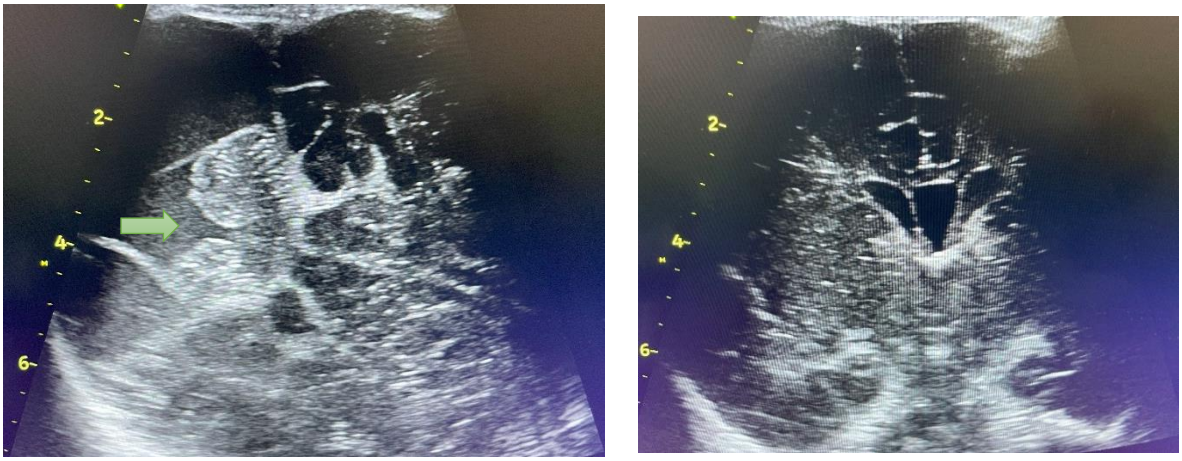

Figure 4: A defect was noted in the roof of the 4th ventricle (Arrow) which is seen communicating with a posterior fossa cyst (Left). The lateral ventricles appear normal. Cavum septum pellucidum is seen (Right).

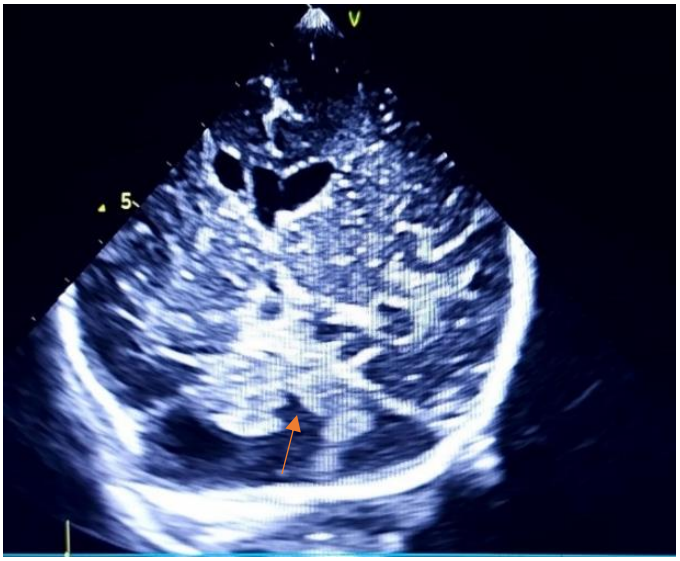

Figure 5: Coronal section at the level of body of lateral ventricles showing the posterior fossa cyst (arrow).

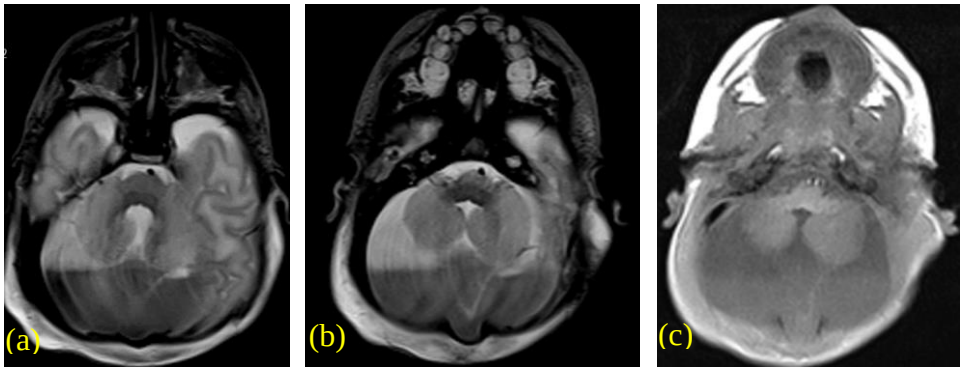

Figure 6: T2WI showing a defect in the roof of the 4th ventricle (a) and a large posterior fossa cyst communicating with the 4th ventricle with blood-CSF level appearing hypointense on T2WI and hyperintense on T1WI (c).

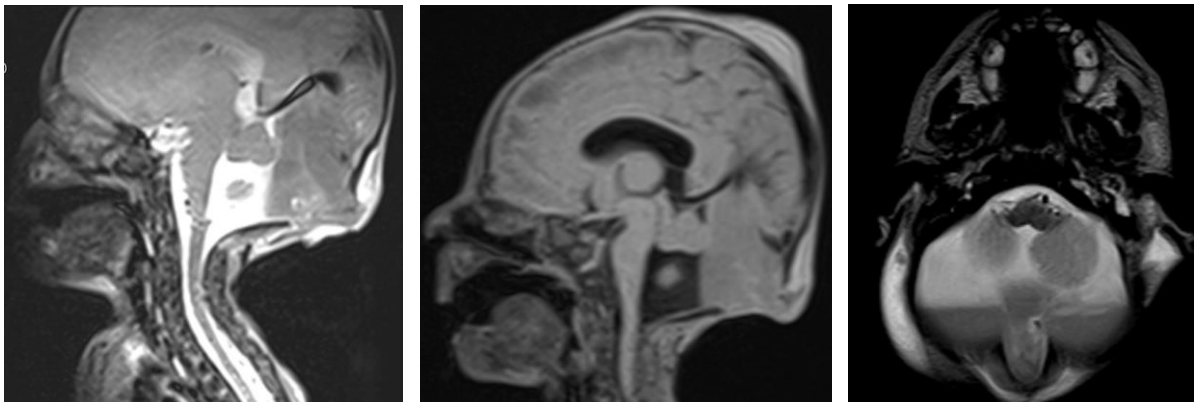

Figure 7: T2WI in sagittal and axial planes reveal a defect in the calvarium in the occipital region across which the posterior fossa cyst is noted to be herniating. The vermis appears hypoplastic and upturned.

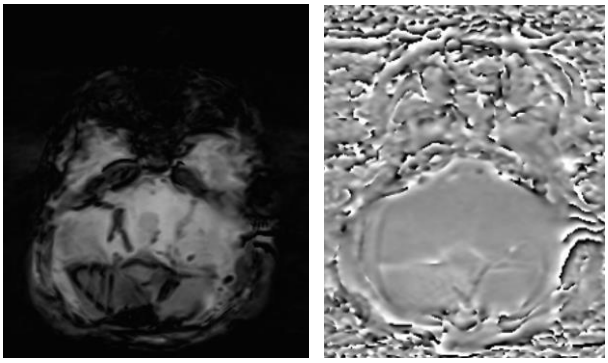

Figure 8: SWI (Left) and phase (Right) images demonstrating the blood-CSF level in the posterior fossa cyst as magnetic susceptibility artefacts appearing hyperintense in phase images

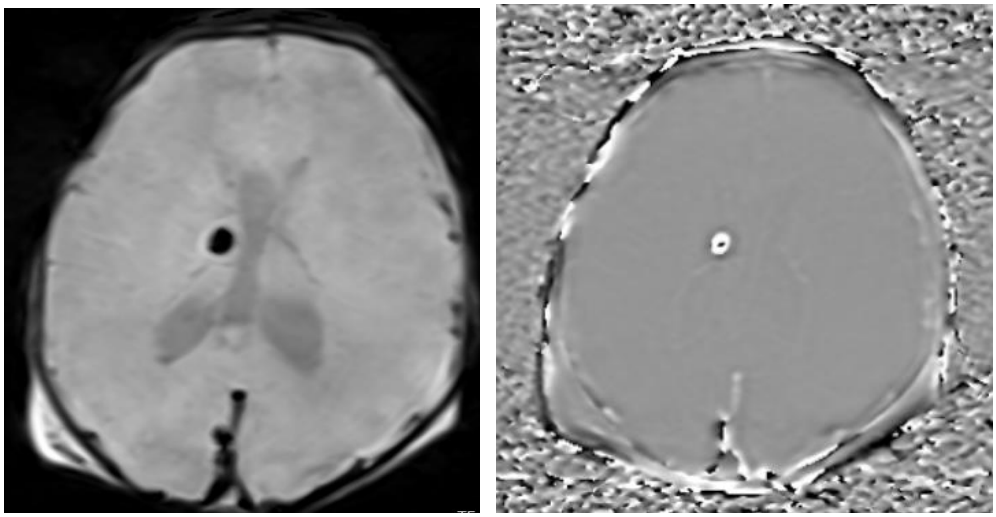

Figure 9: A small focal area of blooming is noted in the right periventricular region

Subgaleal hematoma was noted in bilateral parietal regions, as evidenced by a Subgaleal collection appearing hyperintense on T2WI with attenuation on FLAIR images and magnetic susceptibility artefacts on SWI images, suggestive of birth trauma.

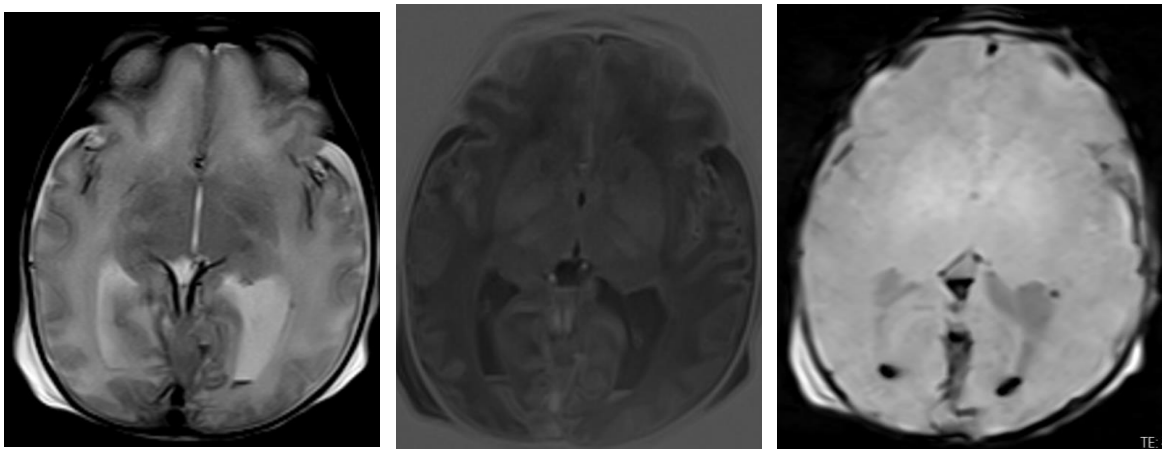

Figure 10: Blood- CSF level is noted in the occipital horns of bilateral lateral ventricles.

DISCUSSION

Abnormal cerebellar development can be either malformations of the paleocerebellum or neocerebellum. Palaeocerebellar malformations typically exhibit vermian defects as key features, whereas malformations of the neocerebellum, including aplasia, hypoplasia, and dysplasia of the cerebellar hemispheres. One such palaeocerebellar developmental anomaly is Dandy walker syndrome.

In 2003, Klein et al. revisited the criteria initially proposed by Barkovich et al., emphasizing the radiological diagnostic criteria for Dandy walker syndrome: [(2)]

- Large median posterior fossa cyst communicating with the fourth ventricle
- Absence of the lower portion of the vermis to varying degrees (lower 3/4, lower half, lower 1/4)
- Hypoplasia, anterior rotation, and upward displacement of the remaining vermis
- Absence or flattening of the fastigium angle
- Large bossing posterior fossa with elevation of the torcular
- Anterolateral displacement of normal or hypoplastic cerebellar hemispheres.

There are various congenital anomalies which share few of the imaging features of the imaging features of the Dandy Walker syndrome and were formerly known as Dandy Walker variants. However, in present times this is being replaced by more specific anatomical nomenclatures.

Embryologically, if the anterior membranous area fails to incorporate into the choroid plexus or there is delay in fenestration of the Foramen of Magendie, then CSF pulsion causes the non-integrated anterior membranous area to balloon posteriorly within the posterior fossa. This cyst does not communicate with the subarachnoid space". [(1)]

It is commonest congenital cerebellar malformation with estimated prevalence of 1:25,000 to 1:50.000 live births. There is female predominance with F:M ratio of 1.5 to 2:1.

Patient may be symptomatic or to have a mild or almost asymptomatic course. In most cases the disturbances are present since birth or in the first year of life. The infants present with hydrocephalus [(3)]. Developmental delay (67%) and epilepsy (32%) are the commonest clinical presentation overall [(1)]. Spastic paraparesis is the most common motor deficit in DWM and DWV disorders, while focal neurological deficits like nystagmus, cranial nerve palsies, truncal ataxia, explosive speech, and dysmetria are relatively rare. These conditions are often associated with extra- and intracranial anomalies, and focal deficits can arise with increased intracranial pressure. Cerebellar deficits typically affect axial movements more than extremities and usually improve with hydrocephalus control. In older children, milder symptoms can include macrocephaly, intracranial hypertension, anomalous movements, ataxia, nystagmus,

headaches, cognitive disabilities, and seizures. It may be associated with non-CNS and chromosomal anomalies.

On imaging, anomalies frequently associated with DWM are ventriculomegaly, hydrocephalus (48%), corpus callosal dysgenesis (10%), encephalocele and various degrees of holoprosencephaly. [(1)] Transcranial ultrasound and MRI can be used for identification of the associated anomalies. In our case, the fetal ultrasound was performed 4 days prior to the delivery which demonstrated Dandy Walker malformation, with occipital meningocele and did not reveal any haemorrhage within the cyst or the occipital meningocele. However, on postnatal imaging, haemorrhage with blood-CSF level was seen.

Haemorrhage within the occipital meningocele seen in the case of interest is rarely reported. Prior case of intrapartum haemorrhage associated with 'acquired dandy walker malformation has been reported in an article by Pichiecchio A et al[(4)]. In this case fetal MRI revealed cerebellar and vermian hypoplasia with 4th ventricular enlargement and cerebellar hemosiderin deposits. This in post-natal scan progressed with superior rotation of the hypoplastic vermis to a Dandy Walker syndrome morphology. A few cases of IVH and EDH have been reported post VP shunting. A case of occipital encephalocele associated with haemorrhage has been reported by D. W. K. Man' and D. M. Forrest in case series of isolated occipital encephalocele. [(5)] However, no case of spontaneous haemorrhage in the posterior fossa cyst were seen.

Hypothesis:

There are a few hypotheses explaining the haemorrhage in the posterior fossa cyst in our case: -

1. Birth trauma- In our case, patient has occipital meningocele which may get compressed between the calvarium and the bony pelvis during the normal vaginal delivery. To substantiate this theory, there are other evidence of birth trauma in form of discoloration of the skin overlying the meningocele and right clavicular fracture. However, no obvious cephalhematoma was seen.
2. Germinal matrix haemorrhage with extension into ventricles and posterior fossa cyst -the neonate was preterm and had a thalamic bleed and dependent blood-CSF level in the lateral ventricle. However, the degree of haemorrhage within the cyst was far exceeding the haemorrhage within the ventricles.
3. Anomalous vessels- The occipital meningocele may have small anomalous vascular channels which led to the haemorrhage within the posterior fossa cyst.

In our case taking the pregnancy till term and elective caesarean section could have addressed the possible causes of haemorrhagic complication in the posterior fossa cyst.

CONCLUSION

Dandy-Walker Malformation (DWM) is an uncommon birth defect impacting the cerebellum and the fourth ventricle's development. In a distinctive case, we noted an infrequent co-occurrence of DWM with occipital meningocele and non-traumatic postnatal intraventricular and posterior fossa cyst haemorrhage. This case underscores the necessity of identification and utilisation of strategies like elective caesarean section and taking the pregnancy till term, to avert trauma and haemorrhage within the cyst.

Declaration by Authors

Ethical Approval: No ethical approval was needed in this case.

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Conflict of Interest: No conflict of interest of any of the authors.

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