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Radiology

THE DANDY WALKER SPECTRUM- A PICTORIAL REVIEW OF MR IMAGING

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ABSTRACT A variety of anomalies are found involving the posterior fossa with Dandy Walker continuum forming a significant chunk of these development disorders. The diagnosis is essential to be done in the antenatal period for timely interventions to be undertaken. However, in a resource poor setup like ours, we find such anomalies quite often escape detection in the intrauterine phase and to know the MRI findings which are apparent in these disorders is quintessential for assessment of the pediatric brain.

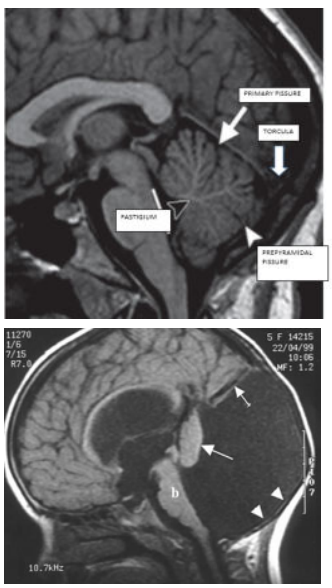
KEYWORDS : Dandy walker, hydrocephalus, Blake's pouch, Mega cisterna magna

INTRODUCTION

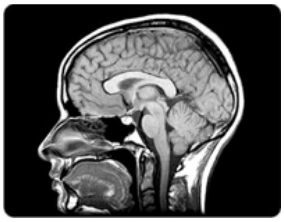
There are numerous developmental defects of the posterior fossa that make up the Dandy Walker continuum. These include Blake's pouch cyst, Dandy Walker variation, and Dandy Walker malformation, which are the most prevalent anomalies in the posterior fossa (inclusion of the fourth ventriculocele in the continuum is controversial).

To correctly diagnose and aid in the pathology's prognostication, their traits must differ just enough from one another. In this article we review the salient findings found in various aspects of this spectrum with few of the images from cases we came across in our institution.

NORMALANATOMY



Measurement of tegmento vermian angle:



Line 1: Drawn along the dorsal surface of the brainstem
Line 2: Drawn along the ventral surface of the vermis

INTERPRETATION:

Normally the angle is close to 0° and an elevated angle is often associated with posterior fossa malformations (8, 9):

- Dandy-Walker malformation : >45°
- vermian hypoplasia : 18-45°
- Blake's pouch cyst : 18-45°
- mega cisterna magna : <18°
- arachnoid cyst : 0°

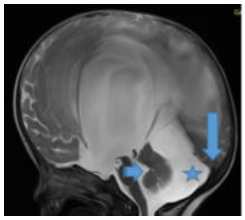
Dandy Walker Malformation (DWM)

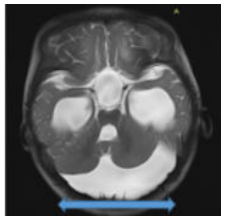
The most frequent posterior fossa malformation, known as DWM, usually happens seldom. The overall risk of recurrence is negligible (1%–5%). (1). DWM can occur alone or in conjunction with other Mendelian disorders like Ritscher-Schinzel syndrome or cranio-cerebello-cardiac syndrome (Online Mendelian Inheritance in Man [OMIM] 220210) or chromosomal anomalies. In some DWM patients, uncommon mutations in six genes (ZIC1, ZIC4, FOXC1, FGF17, LAMC1, and NID1) have also been discovered.

The majority of patients first exhibit signs and symptoms of elevated intracranial pressure during infancy.

The main neuroimaging characteristics of DWM are (a) raised, upward-rotated hypoplasia (or, rarely, agenesis) of the cerebellar vermis (whose inferior section is frequently afflicted, potentially along with its superior portion); and (b) dilatation of the cystic-appearing fourth ventricle, which consequently may fill the entire posterior fossa. Both findings are consistently present and are required for the diagnosis.

Figure 1- In the sagittal T2WI, there is large posterior fossa cyst (★) communicating with an enlarged 4th ventricle (➡). There is torcular inversion (⬇) and gross upstream hydrocephalus. There is gross enlargement of the posterior fossa as appreciated on the axial image.

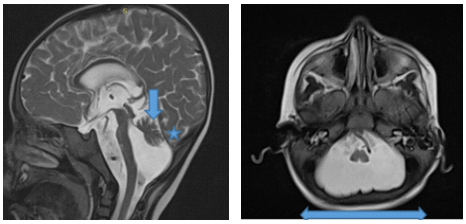




Dandy Walker Variant

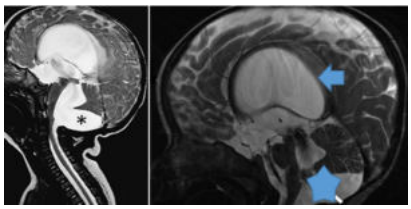
A milder version of the Dandy Walker continuum is the Dandy Walker variation. Vermian hypoplasia and partial fourth ventricle blockage are its defining features. Since the vermis does not develop until after 18 weeks, it cannot be detected on prenatal ultrasound before then.

It is challenging to distinguish between the Dandy-Walker variation and mega-cisterna magna in the pregnancy because clear-cut standards have not been clearly established. When there is a thin link between the fourth ventricle and the cisterna magna, the first condition should be suspected, and the second when the cisterna magna is deeper than 10 mm [2]. Figure 2- On the sagittal T2WI, there is partial vermian hypoplasia (★) along with dilatation of the 4th ventricle. However, there is no upward rotation of the vermis or torcular inversion seen (⬇). Axial images show an enlarged posterior fossa (➖)



Blake's pouch cyst

Insufficient fenestration of the Blake pouch results in a Blake pouch cyst (16–18), which causes tetraventricular hydrocephalus and prevents communication between the fourth ventricle and the subarachnoid space. It symbolises the inferior medullary vellum expanding into the cisterna magna. The displacement of the choroid plexus beneath the vermis along the cyst's anterior and posterior sides is caused by the cyst's presence. Figure 3 - There is an infravermian cyst (✦) which communicates with the dilated 4th ventricle with supratentorial hydrocephalus (⬅). The vermis appears normal (⬟)

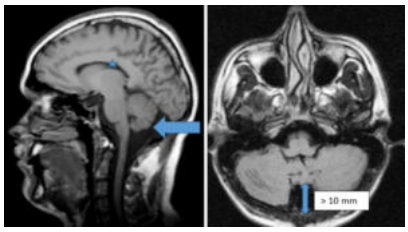


Other posterior fossa abnormalities

Mega cisterna magna

Mega cisterna magna is an enlarged cisterna magna (>10 mm on mid-sagittal imaging) with a normal fourth ventricle, an intact vermis, and, in certain instances, an expanded posterior fossa (4 to 6). Blake's pouch cyst is thought to originate from Blake's pouch absence, while mega cisterna magna is thought to result from Blake pouch's delayed fenestration (5). It can be distinguished from Blake's pouch cyst by the absence of hydrocephalus.

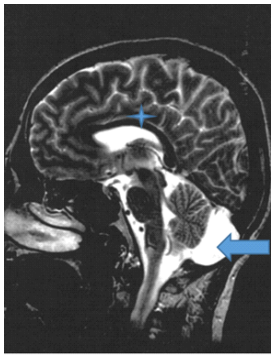
Figure 4- There is a large posterior fossa cyst (⬅). No hydrocephalus can be seen (★).



Arachnoid cyst

Arachnoid cysts are fluid-filled cysts caused by arachnoid membrane duplications. The fourth ventricle or the subarachnoid area are not in communication with arachnoid cysts. There is no known risk of recurrence. Children with posterior fossa arachnoid cysts may exhibit macrocephaly, symptoms of elevated intracranial pressure, and developmental delay, especially if CSF flow is obstructed (7)

Figure 5- There is a posterior fossa cyst posterior to the vermis(➖)with a normal 4th ventricle and supratentorial dilatation of ventricular system (✦).



CONCLUSION

There are multiple conditions resulting in posterior fossa expansion. The differentiation between the conditions needs consideration of subtle elements as well as clincal correaltion and presentation. These congenital conditions are mostly sporadic. Timely diagnosis in antenatal scans can help in their appropriate and timely management. Many conditions are asymptomatic and incidental findings in cranial imaging. The differentiation between these conditions needs a fine eye to the various superimposing features and the subtle clinching factors that help in their identification.

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