Cystic lesion of posterior cranial fossa: is it Dandy-Walker?

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Abstract

Accidental discovery of a fluid collection within the posterior cranial fossa in a fetus or a newborn can be a tricky incidental finding during a routine scan, alarming for a Dandy-Walker Malformation (DWM).

The main cystic lesions of the posterior cranial fossa are DWM, Blake’s Pouch Cyst (BPC), Arachnoid Cyst (AC) and Mega Cisterna Magna (MCM), although the latter is not a proper cyst. The key event for the development of a DWM is a cerebellar vermis hypoplasia that causes the persistence of the superior membranous area, which expands into the posterior fossa forming a large cystic 4th ventricle. BPC is caused by the persistence and herniation of a different membrane, the inferior membranous area, that is supposed to disappear leaving a median opening that would become the foramen of Magendie.

MCM originates if this membrane eventually disappears, leaving an enlarged posterior fossa cavity filled with cerebrospinal fluid physiologically connected with the subarachnoid fluid. Finally, ACs are caused by a defined duplication of the arachnoid membrane filled with CSF-like fluid. Consequently, the radiological finding of a regular cerebellar vermis excludes the hypothesis of DWM and the position of the choroid plexus helps differentiating between DWM and BPC in controversial cases. Moreover, radiological findings in DWM include cystic dilatation of the 4th ventricle and enlargement of the posterior fossa. Absence of hydrocephalus comes out in favor of MCM. Absence of communication with surrounding cerebrospinal fluid defines an AC.

This review assesses the cystic lesions of posterior cranial fossa on the basis of embryological development, radiological findings and associated clinical aspects, in order to clarify the radiological differential diagnosis through embryology.
Keywords

MRI, Sonography, Imaging, Posterior Cranial Fossa, Dandy-Walker Malformation, Subarachnoid Cyst.

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How to cite


Background

Encephalic US scans are now routinely made to point out ischemic suffering of the cerebral parenchyma or signs of infectious disease in case of neonates presenting respiratory distress. We are therefore experiencing an increase of the overall number of scans, performed either by radiologists or clinicians, and that leads to an augmented number of incidental findings. When a fluid collection within the posterior cranial fossa is discovered, the first concern is about a Dandy-Walker Malformation (DWM), that has to be distinguished from other cystic or cystic-like lesions. Although there are several classifications of posterior cranial fossa malformations, we are considering only the ones that primarily comes into differential diagnosis with DWM, which are Blake’s Pouch Cyst (BPC), Arachnoid Cyst (AC) and Mega Cisterna Magna (MCM). This review will discuss those lesions in order to solve some diagnostic doubts, using embryology as a guide to explain the different morphological aspects seen by imaging. The aim is to recall some basic concepts and make them easy to consult, explaining mostly US and MRI findings, that are then examined in depth for a certain differential diagnosis.

Embryology recall

According to the actual knowledge, the development of the cerebellum should be concluded around the 18th week of gestation and pre-natal diagnosis should therefore be avoided until 18/20-week gestation, in order to reduce the number of false positive findings [1, 2]. From that time on, the spot of a cystic dilatation of the posterior cranial fossa should be considered reliable and in need of further investigation. Describing the exact pattern of embryological development of the cerebellum and the 4th ventricle is not the aim of this review, that is meant to be a practical reminder of what’s underneath a fluid collection (Fig. 1 and Fig. 2).

What is important to recall is the pathway that leads to a physiological liquor outflow from the developing 4th ventricle to the subarachnoid space. Although there is still a debate over the timing of developmental events, there is general agreement on their sequence. The cerebellar vermis appears to form by fusion of the cerebellar hemispheres superiorly and medially during the 9th gestational week [3]. During the development of the cerebellum, its ventral portion, which corresponds to the roof of the 4th ventricle, stays almost inactive. Between this portion and the plica chorioidea is located the superior membranous area (SMA), that becomes thicker in a cranio-caudal direction and eventually disappears being included in the midline cerebellar vermis, leaving the choroid plexus attached to the caudal portion of the cerebellum. Caudally to the plica chorioidea, there is another membrane, an ependymal one, named inferior membranous area (IMA), that initially expands forming a recess and eventually disappears leaving a median opening called foramen of Magendie [4]. This event is dated around the 4th month of gestation, whereas the lateral foramina of Luschka form later [5]. The persistence of those membranous areas is the key event for the development of DWM, BPC and MCM and understanding their location in regard to the 4th ventricle helps to quickly distinguish them (Fig. 3 and Fig. 4).

The mechanism of formation of the last cystic lesion that we are considering, the AC, is still debated and so are the causes of cyst enlargement. The most accredited theories involve a primary malformation of the arachnoid membrane or cerebral lobe agenesis and a possible fluid hyper secretion by the lining cells of the cyst wall. Other theories consider a ball-valve mechanism and an osmotic gradient between the intra- and extracystic medium [6]. Regardless the mechanism of formation, which is completely different from the others we have considered, an AC located into the posterior cranial fossa may simulate a pathology involving cerebellum and the 4th ventricle.
development and it is therefore important to consider that into differential diagnosis.

**Differential diagnosis**

In some cases, cystic lesions may be asymptomatic and can be detected incidentally during a scan made for other reasons. In those patients the diagnosis will be often made through a CT scan. When talking about fetuses or babies, CT is considered a not valid diagnostic option because of its radiant dose and must be substituted by ultrasound and MR imaging. Transfontanellar US scans are quick and useful to detect signs of posterior fossa fluid collections and hydrocephalus in newborns and may provide other precious findings in collaborative little patients. It’s hard to make a certain diagnosis using US alone, but it is an important first level study that shouldn’t be forgotten. On the other hand, MRI is surely the most appropriate investigation and permits to identify almost all the findings needed for a correct diagnosis. The only limit is the duration of the scan, that is hard to conciliate with the patience of a newborn, so technically incorrect studies and some movement artefacts must be taken into account.

**Dandy-Walker Malformation**

DWM is the most common posterior cranial fossa malformation. Its prevalence is reported to be 1: 25,000/30,000 and it is mainly sporadic with a low recurrence risk [7, 8].

There’s still a debate over the embryologic development of this malformation, but the most accredited theory points out a cerebellar vermis hypoplasia as the key event. In addition, other observations suggesting that the vermis hypoplasia must be an independent event are that not all the patients with isolated absence of the foramen of Magendie develop a cerebellar
Figure 2. Cerebellar development. A. A bilateral thickening occurs in the alar plate of the rhombencephalon, forming the rhombic lips, which are the primordial cerebellar hemispheres. At 11-13 weeks the transitory external granular layer forms, due to the cellular migration from the germinal zone to the cerebellar surface. B. At 16 weeks, cells from the external granular layer migrate inward to form the internal granular layer of the cerebellar cortex and eventually synapse with cells of the deep cerebellar nuclei. Cells for the deep cerebellar nuclei and the Purkinje layer originate in the wall of the 4th ventricle and migrate directly.

hypoplasia [9] and not all those with DWM present with hydrocephalus [10]. The latter is an unusual event, as the hydrocephalus develops in 70-90% of patients, but there are cases of patients with DWM with a normal fluid outflow allowed by patent foramina of Luschka [11]. A possible range of patency of the foramina and hypoplasia of the vermis may constitute the so-called Dandy-Walker Variant. This name is given to all those syndromes that do not encounter all the characteristics of DWM but still seem to belong to this group of pathologies. Instead of naming it Dandy-Walker Variant, which is an ambiguous definition, a complete anatomical description should be made.

According to this hypothesis, the insufficient of cerebellar vermis development is the cause for the persistence of the SMA that is situated between the cerebellum and the tela choroidea. The usual presentation evidences an atresia of the foramen of Magendie and possibly of the foramina of Luschka. The increase in fluid pressure makes this membrane herniate posteriorly, forming a large cystic 4th ventricle that has no communication with the subarachnoid space, except for the possible presence of the foramina of Luschka.
Those foramina can be patent and allow a partial fluid outflow, that is usually sufficient to avoid the development of in-utero hydrocephalus, but becomes insufficient for a normal fluid outflow in the first month following birth [12].

The radiological triad of signs defining a DWM consists of: vermis hypoplasia, cystic 4th ventricle and enlarged posterior fossa.

**US findings**

When a cystic lesion is detected, it is important to determine whether it is connected with the 4th ventricle or not. Sagittal midline sections are the most useful to obtain a better view of the communication. Using sagittal sections, it is also possible to underline the presence of secondary signs, such as dilatation of third and lateral ventricles or corpus callosum aplasia. Coronal projections are better to evaluate the lateral displacement of the cerebellar hemispheres and the superior displacement of the tentorium [12, 13] (Fig. 5).

**CT findings**

CT scans allow to detect multiple signs of DWM, first of all pointing out a complete or partial aplasia of the cerebellar vermis and the presence of a cystic dilatation of the 4th ventricle. The cerebellar hemispheres appear hypoplastic and laterally displaced, whereas the pons is displaced anteriorly and the imprint of transverse sinuses appears in a higher position. Looking at the bone structure, a bulging and thinning can be detected in the posterior fossa. CT scans are useful to depict a DWM, but for a better morphological study it is mandatory to perform a MRI study [14, 15] (Fig. 6).

**MRI findings**

MRI scans offer a better visualization of the sagittal structures that can be poorly defined in CT scans due to reconstruction. The grade of cerebellar vermis hypoplasia and the absence of the falx cerebelli can be therefore described and the communication between the enlarged posterior fossa and the 4th ventricle visualized. As a consequence of the dilated fossa, the tentorium is displaced superiorly and the lateral sinuses form an inverted “Y” at their junction with the sagittal sinus. Those signs are useful to differentiate a normal but rotated vermis from a hypoplastic one, as the rotation may depend on another cause of increased fluid pressure compressing the vermis. In those cases it is also useful to search for the choroid plexus, that in DWM is localized in a lower position, instead of being attached to the cerebellum. When a proper visualization of the vascular structures is possible,
the absence of the inferior vermian branches and inferior vermian veins can be underlined [16, 17]. MRI is also useful to assess the presence of supratentorial anomalies associated to DWM, therefore improving the prognostic value of the examination, because studies suggest that DWM has a poorer prognosis if associated with supratentorial anomalies [18]. Anomalies including dysgenesis or agenesis of the corpus callosum, occipital encephalocele, polymicrogyria, and heterotopia, may be present in 30-50% of individuals [19] (Fig. 7 and Fig. 8). Nowadays, the duration of the MRI scan is not so long and in many instances images of good quality can be obtained after feeding the baby to avoid movement’s artifacts; for this reason, MRI is currently considered essential in case of cystic lesion of posterior cranial fossa.

**Blake’s Pouch Cyst**

The formation of Blake’s pouch is a physiological step through the embryologic development of the posterior cranial fossa, but its persistence is pathological. This cyst is due to the persistence of the IMA, situated underneath the choroid plexus, that expands backwards instead of reabsorbing.
Figure 5. Dandy-Walker Malformation (DWM): ultrasonography. A. Sagittal section: cystic enlargement of 4th ventricle (arrow). B. Coronal section: vermis hypoplasia (arrow) and dilated ventricles. C. Coronal section: mild enlargement of third (asterisk) and lateral ventricles (arrows). D. Sagittal section: detail of left lateral ventricle.

Figure 6. Dandy-Walker Malformation (DWM). CT scan, sagittal (A), axial (B) and coronal (C) sections: vermian hypoplasia, 4th ventricle enlargement (asterisk, A), brainstem displacement, third (asterisk, B) and lateral ventricles (asterisks, C) enlargement, placement of a shunt catheter into the dilated third ventricle (arrow, A), hypoplastic cerebellar hemispheres.
Figure 7. Dandy-Walker Malformation (DWM): sagittal (A), axial (B), coronal (C) T2-weighted images. The sagittal image shows hypoplastic vermis with verticalization and rotation and elevated tentorial insertion, the posterior fossa is enlarged and filled with CSF, enlarged 4th ventricle, two hypoplastic cerebellar hemispheres, brainstem displacement (arrow, A). The cerebellar falx is normal (arrow, B).

Figure 8. Dandy-Walker Malformation (DWM): sagittal (A), axial (B), coronal (C) T1-weighted images and axial (D) T2-weighted images. The sagittal image shows hypoplastic vermis with verticalization and rotation and elevated tentorial insertion (arrow, A), the posterior fossa is enlarged and filled with a huge CSF cavity, corresponding to an enlarged 4th ventricle. Axial and coronal images show the hypoplastic and dysplastic left cerebellar hemisphere.
to form the foramen of Magendie. As a result, a
dilated fluid collection is formed in the posterior
fossa, freely communicating with the 4th ventricle
and only partially communicating with the
subarachnoid space through the lateral foramina.
Since the foramina of Luschka develop during
the fourth month of gestation, they can’t provide
an adequate fluid outflow in early stages, causing
the pouch to enlarge under fluid pressure.
Hydrocephalus and macrocephaly are therefore
the most common presenting features in the
neonatal period [20]. Hypertension may be present
in case of cyst complication such as hemorrhage
or infection, but the condition may also be
asymptomatic because of the physiological fluid
outflow provided by the foramina of Luschka with
the establishment of a compensation [21].

Suggested radiological findings on fetus US
are a normal anatomy and size of the vermis,
mild/moderate anti-clockwise rotation of the
vermis and a normal size of the cisterna magna
[22]. Pre- and post-natal MRI may confirm the
diagnosis, allowing a better visualization of the
structures and confirming the normal morphology
of vermis and cerebellar hemispheres, even if a
mass effect due to hydrocephalus may be present.
The differential diagnosis with MCM is often
based on the presence of persistent hydrocephalus.
Furthermore, adding contrast allows to identify
the choroid plexus, dislocated upward along the
superior wall of the cystic lesion, in contrast with
the inferior dislocation seen in DWM (Fig. 9).

Mega Cisterna Magna

MCM isn’t a proper cystic lesion but it comes in differential diagnosis because of its
imaging appearance. It is a frequent condition
and accounts for approximatively 54% of cyst-
like posterior fossa malformation [23]. The
anomaly is postulated to be caused by a delayed
permeabilization of the IMA, the same involved
in BPC [24]. When the Blake’s pouch eventually
permeabilises, an enlarged cyst-like area remains,
physiologically connected with the 4th ventricle
and the subarachnoid space. MCM is typically an
incidental finding and it is considered a normal
variant with no need of further investigation
and no recurrence risk. Usually this condition is
completely asymptomatic and hydrocephalus is a
rare complication [25], although studies suggest
that some psychiatric manifestation may be
connected to this condition [26].

Classic radiological aspects include no diffuse
hydrocephalus, normal cerebellar vermis and
physiological fluid outflow, in the presence of an
enlarged fluid cavity in the posterior fossa (> 10
mm on mid-sagittal images) [27] (Fig. 10).

Arachnoid Cyst

AC is a benign fluid collection between the
layers of the arachnoid membrane, that represents
a frequent incidental finding. There are different
theories about the development of this condition
that are still a matter of debate. The most accredited
theories consider it as a primary malformation of
the arachnoid membrane, a cerebral lobe agenesis
or a consequence of a fluid hypersecretion by the
lining cells of the cyst wall. Other theories involve
a ball-valve mechanism or a consequence of an
osmotic gradient between intra and extra cystic
medium [6]. AC have a strong predilection for the
temporal lobes and the middle cranial fossa and

Figure 9. Blake’s Pouch Cyst (BPC): sagittal (A), axial (B), coronal (C) T2-weighted images. There is tetra-ventricular
hydrocephalus. The cerebellar vermis is normally formed and the posterior fossa size is normal. The 4th ventricle is markedly
enlarged and communicates with an infravermian cystic formation that effaces the cisterna magna. The cerebellar falx is
normal (arrow, B).
they rarely localize in the posterior cranial fossa, but when they do it, they need to be distinguished from the other cystic or cystic-like lesions and the development of hydrocephalus is a possibility [28]. Another possible complication of AC is subdural hematoma in case of a cyst rupture [29]. Radiological findings include a normal cerebellar vermis and a fluid collection within the layer of the arachnoid membrane that has no communication with the outer subarachnoid space and shows characteristics compatible with the CSF with all sequences. The cystic walls are often too thin to be visualized with MRI (Fig. 11) and this helps to perform a differential diagnosis with DWM (Fig. 8) or vermian hypoplasia (Fig. 12); in US the differential diagnosis can be challenging.

Conclusions

In case of fluid collection in the posterior cranial fossa, the first thing to be checked for is the cerebellar vermis. If this structure is completely developed, the possibility of a DWM should be excluded and in case of a partial cerebellar hypoplasia found in absence of other characteristic traits of DWM, a complete descriptive diagnosis should be made. It should be therefore avoided to refer to all those anomalies as a “Dandy-Walker Variant”, because this not-specific nomenclature leads to some confusion about the pathology. It is also important to remember some embryological basis concerning the 4th ventricle and cerebellar development, even if there is still a debate over the

Figure 10. Mega Cisterna Magna (MCM): sagittal (A) and axial (B) T2-weighted images demonstrate normal vermis (arrow, B), normal 4th ventricle, scalloping of the occipital bone and absence of hydrocephalus. The cerebellar falx is normal.

Figure 11. Arachnoid Cyst (AC): sagittal (A), coronal (B) T1-weighted images and axial (C), T2-weighted image. Images show a retrocerebellar AC with apparent enlargement of the posterior fossa, scalloping of the occipital bone, mass effect and normal vermis and 4th ventricle (arrow, A).
exact mechanism of formation, in order to avoid the confusion and understand the imaging.

Declaration of interest

The Authors declare that there is no conflict of interest.

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