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Review Articles

Diagnostic imaging of posterior fossa anomalies in the fetus and neonate: part 2, posterior fossa disorders ☆

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ABSTRACT

This second portion of a two-part review illustrates examples of posterior fossa disorders detectable on prenatal ultrasound and MRI, with postnatal or pathology correlation where available. These disorders are discussed in the context of an anatomic classification scheme described in Part 1 of this posterior fossa anomaly review. Assessment of the size and formation of the cerebellar hemispheres and vermis is critical. Diagnoses discussed here include arachnoid cyst, Blake's pouch cyst, Dandy–Walker malformation, vermian agenesis, Joubert syndrome, rhombencephalosynapsis, Chiari II malformation, ischemia, and tumors.

1. Introduction

As discussed in the Part 1 of this two-part review, recognizing a posterior fossa anomaly or acquired disorder is an important role of prenatal imaging. Defining the developmental implications of these disorders then becomes a challenge for the clinical team caring for the pregnant patient and her family. In recent years, changes in the terminology of the different entities affecting the posterior fossa of the fetal brain require that the reader frames an understanding of these disorders in the context of updated information.

In the first part of this review, we describe a diagnostic approach with imaging studies that includes identification of posterior fossa size, presence of abnormal retrocerebellar fluid, and determination of cerebellar size and morphology. This second part of the review illustrates the imaging features of various anomalies of the cerebellum and posterior fossa with prenatal and postnatal correlation.

2. Brief overview of fourth ventricle embryology

The fourth ventricular roof is separated in early development into the anterior and posterior membranous areas, divided by the *plica choroidea*, or primitive choroid plexus. The anterior membranous area will give rise to the cerebellar hemispheres and vermis. The more inferiorly situated posterior membranous area represents the site of the future foramen of Magendie. Nonfenestration of the foramen will cause normal, transient dilatation of the primitive developing fourth ventricle in an inferior and posterior direction, and the appearance of this ballooning vesicle has been termed *a Blake's pouch cyst* (BPC) (discussed in more detail below, Section 3) [1]. If the cyst undergoes subsequent delayed fenestration, a mega cistern magna can result [2]. The cyst can also persist into postnatal life, termed *a persistent Blake's pouch cyst*. If the developmental abnormality is more extensive and involves the *plica choroidea* and anterior membranous area as well, there will be varying degrees of agenesis of the fourth ventricular choroid, vermis, and cerebellar hemispheres [3]. When additionally associated with an enlarged posterior fossa, the classic Dandy–Walker malformation (DWM) is present.

3. Megacisterna magna

Megacisterna magna (MCM) refers to benign enlargement of the subarachnoid spaces of the posterior fossa, with a normally-developed cerebellum. There is characteristic absence of a persistent retrocerebellar cyst and free communication between a normally formed fourth ventricle and the subarachnoid space through a patent median foramen [3]. It is considered to be a normal variant. Prognosis is uniformly excellent in the absence of additional anomalies [3].

Prenatal ultrasound will demonstrate a cisterna magna that measures greater than 1 cm in maximal anteroposterior dimension on a standard oblique axial image through the posterior fossa (Fig. 1a & b).

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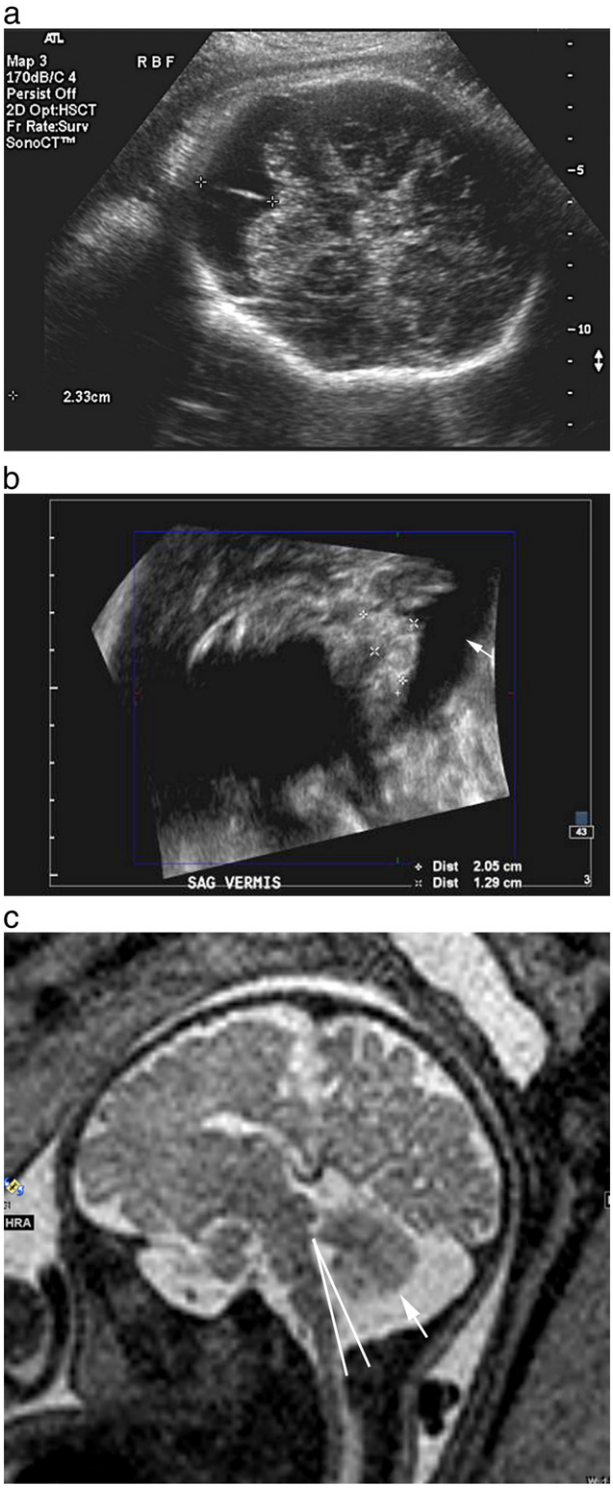

Fig. 1. Mega cisterna magna incidentally detected on prenatal ultrasound. (a) Axial sonographic image from a 36-week prenatal ultrasound demonstrates enlargement of the cisterna magna (2.3 cm marked by calipers) with normal cerebellar contours. Sagittal reformat (b) from a three-dimensional US shows a fully formed vermis and large cisterna (arrow). (c) Midline sagittal T2-weighted image from a prenatal MR at 38-weeks gestational age confirms enlargement of the cisterna magna with normally formed cerebellum and normal tegmento-vermian angle (angle measurement shown in c). Postnatal images confirmed mega cisterna magna in this fetus (not shown here).

The cisterna magna is measured from the posterior margin of the echogenic cerebellar vermis to the inner table of the occipital calvarium [4]. Cerebellar biometry and morphology will be normal, and no evidence of mass effect on the underlying cerebellum should be present [5]. Differential considerations on ultrasound include a persistent BPC, vermian dysplasia, and a DWM.

Fetal magnetic resonance imaging (MRI) of MCM will demonstrate enlargement of the posterior fossa subarachnoid space, similar to that seen on ultrasound (Fig. 1c). Mass effect, as seen with a posterior fossa arachnoid cyst, is absent. A midline sagittal posterior fossa image will characteristically demonstrate a normal contour of the fourth ventricle and cerebellar vermis. In contrast to the malformations detailed in this review, the vermis will not be abnormally elevated away from the brainstem (the tegmento-vermian angle will approach zero), and the vermis will be normal in size [5].

Postnatal ultrasound and MR will demonstrate findings similar to prenatal imaging, with prominence of the cisterna magna. There can be mild associated enlargement of the posterior fossa with subtle scalloping of the occipital calvarium, but marked bony remodeling or mass effect on normal posterior fossa structures suggests an alternate diagnosis. Cerebellar morphology and growth remain normal.

4. Blake's pouch cyst

As described above (Section 1.2), BPC refers to the abnormal persistence of a normal developmental membrane past the 10th gestational week. Lack of normal fenestration of the median foramen results in impaired cerebrospinal fluid (CSF) egress from the ventricular system into the subarachnoid space and the nonfenestrated membrane balloons out into a retrocerebellar cyst (Fig. 2a & b). By definition, the vermis and cerebellar hemispheres are normally formed [2,6]. Depending on the patency of the foramina of Luschka, persistent BPC can therefore present with hydrocephalus of varying severity. Presence of hydrocephalus is the major determinant of prognosis, as this is often an otherwise benign and asymptomatic anomaly.

On prenatal ultrasound, persistent BPC can present with apparent enlargement of the posterior subarachnoid space, mimicking MCM on the standard posterior fossa view. However, an adequate midline sagittal image with either ultrasound or MR can provide characteristic features. These include (a) normal vermian size and morphology; (b) counterclockwise rotation of the vermis with enlargement of the fourth ventricle and increased tegmento-vermian angle; and (c) visualization of the cyst roof at the inferior aspect of the cerebellar vermis. Difficulty in distinguishing inferior vermian compression from partial agenesis can make prenatal distinction between inferior vermian dysplasia with retrocerebellar cyst and persistent BPC difficult or impossible [3]. Use of three-dimensional sonographic technique may allow for more accurate assessment of vermian morphology on reconstructed midline sagittal views and can accentuate subtle differences between the cyst contents and the surrounding subarachnoid CSF in the posterior fossa [2,7,8].

Late fenestration has been documented in up to 60% of cases, and postnatal imaging may demonstrate only a prominent cisterna magna, or may be entirely normal (Fig. 2c) [2]. If present postnatally, BPC will demonstrate identical imaging findings to prenatal imaging. Postnatal distinction of cerebellar vermian dysplasia and BPC is facilitated by high-resolution mid-sagittal imaging demonstrating normal vermian lobulation in the latter [3]. Post contrast T1-weighted MR imaging can also be helpful by delineating the location of the enhancing fourth ventricular choroid plexus: it is deficient in vermian agenesis, but is displaced into the roof of a BPC. The choroid plexus will be normally located in the case of mega cisterna magna and posterior fossa arachnoid cyst [8]. Use of contrast is only appropriate with postnatal imaging, as the risk of fetal neurotoxicity from gadolinium exposure in utero warrants avoiding this contrast agent during fetal MRI.

5. Dandy–Walker malformation

The DWM is believed to result from failure of normal development of the membranous roof of the primitive fourth ventricle [1,3,9]. The DWM consists of partial agenesis or absence of the cerebellar vermis