MRI characteristics of posterior cranial fossa malformations.

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Learning objectives

A wide spectrum of congenital anomalies affects posterior cranial fossa structures. The purpose of this exhibit is to demonstrate developmental pathologies of the posterior cranial fossa.

Background

By the 4th gestational week fusion of the neural tube in the cranial region and closure of the rostral neuropore forms the three primary brain vesicles: forebrain (prosencephalon), midbrain (mesencephalon), and the hindbrain (rhombencephalon).

Posterior fossa develops mainly from hindbrain which further divides into two secondary vesicles: the metencephalon and the myelencephalon. The pons and the cerebellar hemispheres develop from the metencephalon. The medulla oblongata - from the myelencephalon. The cavity of the hindbrain becomes the future 4th ventricle. The vermis takes its origin from the midbrain (Fig. 1)

Images for this section:
Fig. 1: Diagram illustrating five-vesicles stage of the developing brain.
Findings and procedure details

1. Malformations of both midbrain and hindbrain

Brainstem-cerebellar hypoplasia-dysplasia. This group of malformations includes a wide spectrum of disorders from the very rare cerebellar agenesis to more common hypoplasia with diffuse brainstem involvement [1] (Figs. 2-4).

Chiari II malformations are encountered relatively commonly. This condition is characterised by a small posterior fossa with descent of its structures into the spinal canal through the wide foramen magnum and by - usually lumbosacral - spina bifida aperta/myelomeningocele (Arnold-Chiari malformation) (Figs. 5-6). Numerous associated abnormalities are also frequently encountered, e.g. callosal hypoplasia/dysgenesis, stenogyria [2] (Fig. 7). In Chiari III malformation Chiari II signs are present and additionally cervico-occipital encephalocele with hindbrain dysplasia is seen [2] (Figs. 8-9).

Cobblestone lissencephaly - reduction in normal sulcation, associated with a bumpy or pebbly cortical surface as a result of overmigration and numerous additional posterior fossa anomalies: hypoplastic and dysmorphic brainstem with a posterior kink and hypoplastic cerebellum. A number of different genetic abnormalities have been identified resulting in cobblestone lissencephaly. One of the most commonly included syndrome is Walker-Warburg sy [2][3] (Figs. 10-12).

Molar tooth sign-associated malformations (i.e. Joubert sy and associated disorders). Molar tooth sign results from a midbrain-hindbrain malformation characterized by thickened and elongated superior cerebellar peduncles, hypoplastic vermis and an abnormally deep interpeduncular fossa. One of the known syndromes associated with molar tooth sign is Joubert sy. In this condition variable degree of cerebellar vermian agenesis is present. Brainstem dysplasia is also common [4] (Figs. 13-15). Joubert syndrome related disorders include - among others - orofacial-digital sy type VI [5].

Rhombencephalosynapsis (RES) is characterised by partial or complete absence of the cerebellar vermis with continuity of the hemispheres across the midline. The cerebellar hemispheres, peduncles and dentate nuclei are fused. RES is often associated with midbrain abnormalities (aqueductal stenosis and midline fusion of the colliculi) and supratentorial abnormalities [6] (Figs. 16-18).

2. Malformations affecting mainly cerebellum and its derivatives
Focal or hemispheric hypoplasia- a broad group of malformations characterised by hypoplastic cerebellar structures observed focally [1] (Figs. 19-20).

Paleocerebellar hypoplasia is characterised by defective development of the most of the vermis and of the adjacent zones of the cerebellar hemispheres rostral to the primary fissure as it is in Dandy-Walker malformation (DWM) and cerebellar vermis hypoplasias. DWM comprises the triad of agenesis or hypoplasia of the vermis with its cephalad rotation, cystic dilatation of the fourth ventricle and enlarged posterior fossa due to elevated tentorium [1][7] (Figs. 21-23).

Cerebellar vermis hypoplasia is associated with normal position or minimal upward rotation of the hypoplastic vermis relative to the brainstem. 4th ventricle is only mildly enlarged and the tentorium cerebelli is not elevated [1] (Fig. 24).

Neocerebellar hypoplasia- in this condition hemispheres are predominantly affected [1] (Figs. 25-27).

3. Malformations affecting mainly lower hindbrain

Chiari I malformation is characterised by inferior herniation of the cerebellar tonsils through the foramen magnum, due essentially to a mismatch between the size and content of the posterior fossa (Fig. 28).

Cranial nerves and nuclear aplasias as it is in Möbius syndrome- a rare congential condition characterised by the absence or under-development of the abducens nerve (CN VI) and facial nerve (CN VII) nuclei [1].

4. Developmental abnormalities of the posterior cranial fossa

Abnormal fluid collections

- arachnoid cysts - filled with CSF, do not communicate with the surrounding subarachnoid space and ventricular system; they usually are not associated with brain maldevelopment (Figs. 29-31).

- Blake pouch cyst ( = retrocerebellar arachnoid cyst) - a cystic collection, inferior and posterior to the cerebellum with tetra-ventricular hydrocephalus. In this condition mass effect on the cerebellum is common [7].
-mega cisterna magna - the appearance is similar to a persistent Blake’s pouch except for the consistent absence of hydrocephalus [7] (Figs. 32-34).

Abnormal bone and brain structure

The example of this group of malformations may be hemimegalencephaly with ispilaterally altered gyration of the cerebellum (Figs. 35-36)

5. Malformations associated with prenatal onset degeneration

Pontocerebellar hypoplasia - typically MRI demonstrates a progressive atrophy of the ventral pons and the inferior olivary nuclei, vermis and cerebellar hemispheres [1].

Congenital disorders of glycosylation are genetically heterogeneous autosomal recessive disorders caused by abnormal glycosylation of N-linked oligosaccharides. Cerebellar volume loss (cerebellar hypoplasia or pontocerebellar atrophy) at presentation and on follow-up is the most characteristic feature of these group of disorders. Diminution of the brainstem and cerebellar T2/FLAIR hyperintensity may also be seen [8].

6. Other malformations and syndromes

Lhermitte-Duclos syndrome results from derangement of normal laminar cellular organization of cerebellum. Widened cerebellar folia with a striated appearance on MRI is usually confined to one hemisphere, occasionally extended to the vermis and only rarely to the contralateral hemisphere [5][9].

Macrocerebellum (in Alexander disease). In these condition diffusely enlarged cerebellum may be associated with delayed white matter myelination. In Alexander disease MR imaging also shows extensive cerebral white matter abnormalities with a frontal predominance and brain stem abnormalities, in particular involving the midbrain and medulla [10].

Aicardi syndrome - rare neurodevelopmental disorder with the high incidence of cerebellar abnormalities: superior prominence of the vermis and its inferior hypoplasia, dysplasia, hypoplastic hemispheres, enlarged cisterna magna and cerebellar cysts [5].

Smith-Lemli-Opitz syndrome is an autosomal recessive disorder characterised by multiple developmental malformations including cerebellar hypoplasia [5].
7. Additionally we would like to present unusual inborn infratentorial abnormalities combined with supratentorial malformations that we are not sure where to place in the existing classifications. A 15-month-old boy was hospitalized and diagnosed because of epilepsy. The clinical diagnosis was cerebral palsy.

Magnetic resonance imaging (MRI) revealed bilateral polymicrogyria/pachygyria involving most of the frontal and parietal lobes, partially also temporal. Only occipital lobes were spared (Figs. 37-40)

The corpus callosum was underdeveloped in its posterior part, lacking the isthmus. The tegmentum was thickened. There was a molar tooth sign in the infratentorial compartment: deep interpeduncular fossa, thick, elongated superior cerebellar peduncles, cerebellar vermis hypoplasia. The pons was flattened, with decreased anterior-posterior dimension and increased lateral dimension (Figs. 41-44). The flat ventral pons was a feature similar to the cases of pontine tegmental cap dysplasia (PTCD) (Figs. 45-48) but vaulted pontine tegmentum was absent and middle cerebellar peduncles were present. Thickened medulla oblongata (abnormal rounded cap over it) and abnormal anterior outline of the pons make this case similar to medullary cap dysplasia. The thickened tegmentum is found in muscle-eye-brain disease and Walker-Warburg syndrome. Anyway, the malformation must belong to the so-called dystroglycanopathies which are believed to be caused by impaired O-mannosylation of alfa-dystroglycan. In this group of malformations the brainstem is affected in nearly all patients, with enlarged quadrigeminal plates, fusion of the colliculi, hypoplasia of the pons, often with a longitudinal ventral midline pontine cleft. The disorder seems to result from both abnormal neuronal migration and abnormal formation of white matter tracts [11].

Images for this section:
Fig. 2: Prenatal findings of cerebellar and brainstem hypoplasia. SSFSE/T2WI, sag.
Fig. 3: Postnatal examination of the same baby as in Fig. 2 confirms prenatal diagnosis. FSE/T2WI, sag.
Fig. 4: Postnatal examination of the same baby as in Fig. 2 confirms prenatal diagnosis. FSE/T2WI, cor.
**Fig. 5:** Classical appearance of Arnold-Chiari malformation. Small posterior fossa with partial descent of the cerebellum into the spinal canal and small, flattened 4th ventricle. The cisterna magna is obliterated. FSE/T2WI, sag.
Fig. 6: Axial image at the level of the foramen magnum of the same child as in Fig. 5 shows downward migration of the cerebellum which, wrapped tightly around the brain stem, has the appearance of a banana (banana sign) FSE/T2WI, ax.
Fig. 7: An 11-year-old patient operated in the past due to myelomeningocoele. Posterior fossa shows features of Arnold-Chiari malformation. In addition callosal dysgenesis and stenogyria are seen. FSE/T2WI, sag.
**Fig. 8:** An examination of a newborn girl with occipital encephalocele. There is almost complete agenesis of the cerebellum and dorsal kinking of the brainstem which enters the ring of the hernia. Syringomyelia is also seen. FSE/T2WI, sag.
Fig. 9: An examination of a newborn girl with occipital encephalocele. There is almost complete agenesis of the cerebellum and dorsal kinking of the brainstem which enters the ring of the hernia. Syringomyelia is also seen. F SE/T2WI, sag.
Fig. 10: Prenatal examination performed at the gestational age of 22 weeks due to suspicion of vermian hypoplasia revealed hydrocephalus only in the supratentorial compartment (lissencephaly is physiologic at this gestational age so the brain surface could not have been diagnosed as abnormal). The main findings were infratentorial: flattened ventral surface of the pons, thickened tectum, kinking of the brainstem. These findings made a diagnosis of Walker-Warburg sy most likely and it was confirmed after birth. SSFSE/T2WI, sag.
Fig. 11: Prenatal examination performed at the gestational age of 22 weeks due to suspicion of vermian hypoplasia revealed hydrocephalus only in the supratentorial compartment (lissencephaly is physiologic at this gestational age so the brain surface could not have been diagnosed as abnormal). The main findings were infratentorial: flattened ventral surface of the pons, thickened tectum, kinking of the brainstem. These findings made a diagnosis of Walker-Warburg sy most likely and it was confirmed after birth. SSFSE/T2WI, cor.
Fig. 12: Prenatal examination performed at the gestational age of 22 weeks due to suspicion of vermian hypoplasia revealed hydrocephalus only in the supratentorial compartment (lissencephaly is physiologic at this gestational age so the brain surface could not have been diagnosed as abnormal). The main findings were infratentorial: flattened ventral surface of the pons, thickened tectum, kinking of the brainstem. These
findings made a diagnosis of Walker-Warburg syndrome most likely and it was confirmed after birth. SSFSE/T2WI, ax.
**Fig. 13:** An examination of a 4-year old girl with Joubert sy shows molar tooth sign. Cerebellar dysgenesis and hydrocephalus is also seen. SE/T1WI, ax.

**Fig. 14:** An examination of a 4-year old girl with Joubert sy shows molar tooth sign. Cerebellar dysgenesis and hydrocephalus is also seen. FSE/T2WI, ax.
Fig. 15: An examination of a 4-year old girl with Joubert sy shows molar tooth sign. Cerebellar dysgenesis and hydrocephalus is also seen. The shape of the 4th ventricle resembles "bat wings". SE/T1WI, ax.
**Fig. 16:** Examination of a 3-day-old baby with severe hydrocephalus reveals small cerebellum, absence of the cerebellar vermis with fusion of the cerebellar hemispheres. FSE/T2WI, sag.
Fig. 17: Examination of a 3-day-old baby with severe hydrocephalus reveals small cerebellum, absence of the cerebellar vermis with fusion of the cerebellar hemispheres. FSE/T2WI, cor.
**Fig. 18:** Examination of a 3-day-old baby with severe hydrocephalus reveals small cerebellum, absence of the cerebellar vermis with fusion of the cerebellar hemispheres. FSE/T2WI, ax.
Fig. 19: A 3-year-old boy with oblique facial cleft. MR examination shows absence of the left cerebellar hemisphere. FSPGR, 3D/T1WI, cor.
**Fig. 20:** A 3-year-old boy with oblique facial cleft. MR examination shows absence of the left cerebellar hemisphere. FSE/T2WI, ax.
Fig. 21: Examination of a 2-month-old girl revealed a classic radiological picture of Dandy-Walker syndrome. FSE/T2WI, sag.
Fig. 22: Examination of a 2-month-old girl revealed a classic radiological picture of Dandy-Walker syndrome. FSPGR, 3D/T1WI, cor.
Fig. 23: Examination of a 2-month-old girl revealed a classic radiological picture of Dandy-Walker syndrome. FSE/T2WI, ax.

Fig. 24: In this case the vermis is hypoplastic and the 4th ventricle is enlarged but the tentorium is not elevated and posterior fossa volume is not enlarged. FSE/T2WI, sag.
**Fig. 25:** A 7-year-old boy with a positive family history of encephalopathy. Diffuse neocerebellar hypoplasia is seen. FSE/T2WI, sag.
**Fig. 26:** A 7-year-old boy with a positive family history of encephalopathy. Diffuse neocerebellar hypoplasia is seen. FLAIR, ax.
Fig. 27: A 7-year-old boy with a positive family history of encephalopathy. Diffuse neocerebellar hypoplasia is seen. FSE/T2WI, cor.
Fig. 28: A 6-year-old boy with autism. The tonsils protrude through the foramen magnum by more than 5 mm without any other cerebral changes. FSE/T2WI, sag.
Fig. 29: In this case there is a posterior fossa arachnoid cyst localised behind and under the left cerebellar hemisphere. Thinning and scalloping of the occipital bone is seen. FSE/T2WI, ax.
**Fig. 30:** In this case there is a posterior fossa arachnoid cyst localised behind and under the left cerebellar hemisphere. Thinning and scalloping of the occipital bone is seen. FSE/T2WI, cor.
**Fig. 31:** In this case there is a posterior fossa arachnoid cyst localised behind and under the left cerebellar hemisphere. Thinning and scalloping of the occipital bone is seen. FSE/T2WI, sag.
Fig. 32: This examination shows a cystic collection inferior and posterior to the cerebellum which, with absence of hydrocephalus and of occipital bone modelling - it represents mega cisterna magna. FSE/T2WI, sag.
Fig. 33: This examination shows a cystic collection inferior and posterior to the cerebellum which, with absence of hydrocephalus and of occipital bone modelling - it represents mega cisterna magna. FSE/T2WI, ax.
**Fig. 34:** This examination shows a cystic collection inferior and posterior to the cerebellum which, with absence of hydrocephalus and of occipital bone modelling - it represents mega cisterna magna. FSE/T2WI, cor.
Fig. 35: Examination of a patient with right-sided hemimegalencephaly reveals in addition ipsilateral altered gyration of the cerebellum., FSE/T2, cor.
Fig. 36: Examination of a patient with right-sided hemimegalencephaly reveals in addition ipsilateral altered gyration of the cerebellum. FSE/T2, ax.

Fig. 37: Cortical malformations in a 15-month-old boy diagnosed because of cerebral palsy and epilepsy. FSE/T2WI, ax.
Fig. 38: Cortical malformations in a 15-month-old boy diagnosed because of cerebral palsy and epilepsy. SE/T2WI, ax.
Fig. 39: Cortical malformations in a 15-month-old boy diagnosed because of cerebral palsy and epilepsy. SE/T1WI, ax.
Fig. 40: Cortical malformations in a 15-month-old boy diagnosed because of cerebral palsy and epilepsy. SE/T1WI, ax.
**Fig. 41:** The same baby as in figs. 37-40. Posterior fossa and corpus callosum findings. SE/T1WI, ax.
Fig. 42: The same baby as in figs. 37-40. Posterior fossa and corpus callosum findings. FSE/T2WI, ax.
Fig. 43: The same baby as in figs. 37-40. Posterior fossa and corpus callosum findings. FSE/T2WI, sag.
Fig. 44: The same baby as in figs. 37-40. Posterior fossa and corpus callosum findings. FSE/T2WI, ax.
Fig. 45: A 13-month-old baby with developmental delay, generalized hypotonia, vertical pendular nystagmus, and cranial nerves IV, V, and VII paresis. A dorsal vault projecting from the pontine tegmentum into the fourth ventricle. FSE/T2, sag.
Fig. 46: A 13-month-old baby with developmental delay, generalized hypotonia, vertical pendular nystagmus, and cranial nerves IV, V, and VII paresis. Abnormal shape and course of superior cerebellar peduncles. FSE/T2, ax.
Fig. 47: A 13-month-old baby with developmental delay, generalized hypotonia, vertical pendular nystagmus, and cranial nerves IV, V, and VII paresis. Abnormal shape and course of superior cerebellar peduncles. FSE/T2, cor.
**Fig. 48:** A 13-month-old baby with developmental delay, generalized hypotonia, vertical pendular nystagmus, and cranial nerves IV, V, and VII paresis. Very thin/not visible middle cerebellar peduncles. FSE/T2, ax.
Conclusion

MRI is the best method of imaging the posterior cranial fossa structures and demonstrates high efficiency in diagnosis of its malformations. The knowledge of entities which can be expected and diagnostic experience prevent from making a wrong diagnosis.

Personal information

References


