A RARE CASE REPORT OF DANDY WALKER MALFORMATION WITH LISSENCEPHALY, PARTIAL CORPUS CALLOSAL AGENESIS AND HYDROCEPHALUS

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ABSTRACT

Dandy-Walker malformation is characterised by agenesis or hypoplasia of the cerebellar vermis, cystic dilatation of the fourth ventricle, and enlargement of the posterior fossa. Approximately 70-90% of patients have hydrocephalus. Lissencephaly is characterised by absence (agyria) or incomplete development (pachygyria) of the gyri of the cerebral cortex, causing the brain's surface to appear unusually smooth. Our case demonstrates Dandy Walker malformation with partial corpus callosal agenesis, hydrocephalus and Lissencephaly.

Keywords: Dandy Walker malformation, Lissencephaly

INTRODUCTION

Dandy-Walker malformation is a rare abnormality of the central nervous system with incidence of 1 in 25-35,000 live births. Dandy walker malformation consist of hydrocephalus, posterior fossa cyst, agenesis or hypoplasia of cerebellar vermis and other concomitant problems.

CASE REPORT

A thirty five year old female came to our hospital in labor pains and delivered a baby boy. No prior antenatal ultrasound was done. The baby did not cry at birth and was not able to suck milk. Neurosonogram was ordered. It showed gross hydrocephalus with thinned overlying brain cortex. The fourth ventricle was dilated and was seen communicating with a large posterior fossa cyst. MRI was obtained which showed large posterior fossa cyst communicating with the fourth ventricle. There was aplasia of cerebellar vermis. The cerebellar hemispheres appeared displaced laterally by the cyst. The cerebellar hemispheres and brain stem were mildly hypoplastic. Gross hydrocephalus was noted. Partial agenesis of corpus callosum was noted affecting genu and the body. The brain parenchyma appeared thinned and showed pachygyria (incomplete development of gyri) with shallow sylvian fissure. The diagnosis of Dandy Walker malformation with partial corpus callosal agenesis, Lissencephaly and hydrocephalus was done. An attempt was made to drain the dilated ventricles however the baby died on the third day.

DISCUSSION

Posterior fossa cystic malformations are divided into Dandy-Walker malformation, Dandy-Walker variant, mega cisterna magna, and posterior fossa arachnoid cyst. They except posterior fossa arachnoid cyst are believed to represent a continuum of developmental anomalies and consist of the spectrum called as the Dandy-Walker complex [8, 9, 10]. Common findings of Dandy-Walker malformation are: Enlarged posterior fossa with cyst formation, varying degrees of cerebellar and vermian hypoplasia or complete vermian absence, hypoplastic cerebellar hemispheres winged anterolaterally in front of the cyst, absence of the foramina of Luschka and Magendie which is common cause for obstructive hydrocephalus (70 – 90 %), abnormally high position of the straight sinus.
torcular herophili, and tentorium, callosal agenesis (20-25%), brainstem compression and hypoplasia. It may be associated with other nervous system abnormalities like: Dysgenesis of the corpus callosum (20-25%), Holoprosencephaly (25%), Dysplasia of the cingulate gyrus (25%), Polymicrogyria/gray matter heterotopia (5-10%), Occipital encephalocele (7%) and Malformation of the cerebellar folia (25%). Non CNS abnormalities are noted in 25 to 33 percent children. They are: Orofacial deformities and cleft palate, Polydactyly and syndactyly, Cardiac anomalies, polycystic kidneys, Cataracts, retinal dysgenesis, and choroid coloboma.

Ultrasound is the initial examination performed because of its easy availability and as it can be done portably as well as without sedation.[11, 12] Abnormalities such as the gyral, dural, tentorial, and skull anomalies may not be easily picked up on ultrasound study. MRI is considered best imaging modality to define the relationship between the cyst and the fourth ventricle, vermian dysgenesis and associated complications. Lissencephaly literally means smooth brain. It is a disorder caused by defective neuronal migration during the 12th to 24th weeks of gestation resulting in a lack of development of sulci and gyri. MRI is the preferred modality to pick up this abnormality.

CONCLUSION

Dandy Walker malformation with partial corpus callosal agenesis, Lissencephaly and hydrocephalus is one of the rare combinations of congenital defects seen. Even though Ultrasound is primary modality of screening, MRI is preferred to get complete details on the malformation and its complications.

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REFERENCES


Figure 1: Nerosonogram coronal image shows dilated fourth ventricle (*) communicating with the posterior fossa cyst.

Figure 2: Coronal T2W MRI shows dilated fourth ventricle (*) communicating with the posterior fossa cyst. Dilated lateral ventricles and antero-laterally displaced mildly hypoplastic cerebellar hemispheres are seen.
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Figure 3: Axial T1W MRI shows dilated lateral ventricles. Pachygyria is appreciated as well.

Figure 4: Axial T1W MRI shows dilated third ventricle (*) communicating with interhemispheric fissure with colpocephaly.

Figure 5: Sagittal T1W MRI shows enlarged posterior fossa with a large posterior fossa cyst. Vermian aplasia is seen with mild hypoplasia of cerebellar hemispheres and brain stem.