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Case Report

Dandy Walker variant mimicking as cerebral palsy with severe neurological impairment

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ABSTRACT

Dandy Walker Variant (DWV) is a less severe form of the spectrum of Dandy Walker Malformation (DWM). The DWM is a rare congenital intracranial posterior fossa malformation comprising cystic dilatation of the fourth ventricle, complete or partial agenesis of the cerebellar vermis and an enlarged posterior fossa. Clinically it presents with variable degrees of neurological impairment. Definitive diagnosis of DWM or DWV depends on neuroimaging as most of the clinical signs are not conclusive. This child was clinically suspected as spastic quadriplegic cerebral palsy had incidental finding of DWV on neuroimaging. Hence a definitive diagnosis of DWV was made by MRI in this case. Here we discuss the clinical and radiological aspects of this case with DWV without other associated anomalies yet presenting with clinically significant neurological impairment.

Keywords: Dandy Walker malformation, Dandy Walker variant, MRI, Neurological impairment

INTRODUCTION

The Dandy-Walker malformation is a congenital intracranial posterior fossa malformation typically involving fourth ventricle and the cerebellum. The DWM has an estimated prevalence of about 1:25-30000 live births with a slight female predominance[1-3] with male: female ratio of 1:3.[4]

DWM is of multifactorial aetiology but genetic factors have a major role as DWM may occur as part of Mendelian disorders and chromosomal aberrations. Antenatal environmental factors like viral infections, alcohol and diabetes have also been suggested to play a role in the genesis of DWM. In the absence of a detectable syndrome a recurrence risk in subsequent pregnancies of 1-5% is suggested.[5] It also contributes for 1-4% of cases of foetal hydrocephalus on antenatal ultrasound.[6] This child had more severe neurological impairment for her MRI diagnosis of DWV inspite of the absence of any additional intra or extracranial abnormalities.

CASE REPORT

One and a half year old female child 1st child born to non-consanguineous parents presented with convulsions and delay in achieving milestones noticed by parents since seven months of age. Convulsions were generalised tonic clonic with uprolling of eyes, occurring once in every two to three days. Developmentally child had achieved partial head holding and cooing. Child was delivered normally at term to antenatally registered, immunised mother with regular antenatal evaluation and follow up. Baby did not have birth asphyxia or NICU stay and the birth weight was 2700 grams. Antenatal and postnatal period was

uneventful. Child was immunised completely and weight and height were normal for the age.

On examination child was conscious, lying still, had lack of interaction with surrounding and had less movements of both upper and lower limbs. Child had microcephaly and the head circumference was 39 cm for expected of 47 cm, anterior fontanelle was closed, occiput was flattened (brachycephaly) and spine was normal. On neurological examination cranial nerves were apparently normal. Tone was increased in both upper and lower limbs, deep tendon reflexes were brisk and bilateral plantars were extensors. Other systems on examination were normal. Based on the history and examination findings a clinical diagnosis of quadriplegic spastic cerebral palsy with epilepsy was made. EEG done previously was suggestive of generalised epileptiform discharges with basal suppression. MRI Brain detected vermian hypoplasia with non-visualisation of inferior vermis. There was mild dilatation of fourth ventricle which was communicating with cistern magna, suggestive of Dandy Walker variant. Thus MRI brain assigned a definitive diagnosis of Dandy Walker variant in this case.

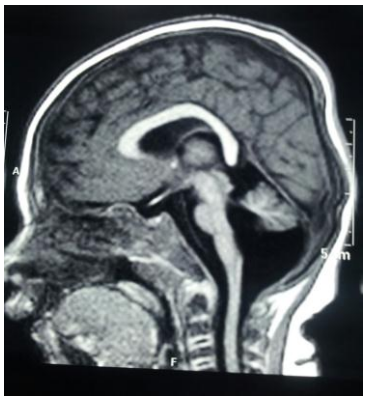

Figure 1: Sagittal image of MRI Brain showing cerebellar hypoplasia with mild dilatation of fourth ventricle communicating with cistern magna.

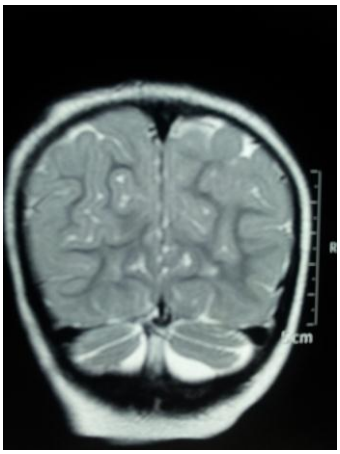

Figure 2: Coronal image of MRI Brain showing hypoplastic cerebellar hemispheres, vermis and non-visualisation of inferior vermis.

DISCUSSION

Dandy-Walker Malformation (DWM) is one of the commonest intracranial congenital malformations affecting the posterior fossa. DWM constitutes a triad of cystic dilatation of the 4th ventricle, complete or partial agenesis of the cerebellar vermis and an enlarged posterior fossa.[7] Dandy-Walker Variant (DWV) is its less severe form, having cystic posterior fossa with variable hypoplasia of the cerebellar vermis and without enlargement of the posterior fossa. Megacisterna magna is presence of enlarged cisterna magna with normal cerebellar vermis and fourth ventricle. All the above spectrum of posterior fossa and cerebellar malformation is referred to as Dandy Walker complex.[8,9] Agenesis of the corpus callosum, holoprosencephaly, occipital encephaloceles and ocular abnormalities frequently coexist with DWM. Extra-cranial anomalies usually associated with DWM are polycystic kidneys, cardiovascular defects, polydactyly and cleft palate.[5]

Children with DWM present with developmental delay, enlarged head circumference, or signs and symptoms of hydrocephalus. Difficulty with balance, spasticity, and poor fine motor control are common. The degree of developmental delay appears to be related to the level of hydrocephalus and to the extent of supratentorial anomalies Seizures occur in 15-30% of patients. Subnormal intelligence is manifested in 41-71% of patients and more severe intellectual impairment occurs in patients with agenesis of the corpus callosum.[10]

DWM has a wide range of clinical spectrum with varying degree of neurological impairment. Dandy Walker Variant (DWV) is a less severe form of the spectrum of Dandy Walker Malformation (DWM). The isolated Dandy Walker variant abnormality has the highest incidence of survival, and there are reported cases of people who have had Dandy Walker variant their entire lives without any symptoms. Patients with Dandy-Walker variant are more likely to present in adulthood than in infancy or childhood.[11]

In the neuroradiological literature a distinction is often made between Dandy-Walker malformation and Dandy-Walker variant; the latter term is applied if the posterior fossa is not enlarged, the hypoplasia of the cerebellar vermis is less pronounced, or both.[12] The introduction of modern imaging techniques, specifically MRI, has radically changed the evaluation of symptoms related to the posterior fossa. MRI usually is performed for detailed evaluation of Dandy-Walker malformation lesions and complications.[4,13]

Our patient had absence of obvious external congenital malformation, had brachycephaly instead of occipital prominence, had microcephaly instead of macrocephaly which were suggestive of milder spectrum of DWM i.e. DWV. But contrary to the above examination findings child had severe neurological impairment involving white

and grey matter manifesting as significant motor delay and seizures. The definitive diagnosis, though an incidental finding was due to advanced neuroimaging technique like MRI.

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