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Original Research Paper

Radiodiagnosis

RADIOLOGICAL EVALUATION OF A RARE CASE OF DANDY-WALKER MALFORMATION IN A 25 YEARS OLD MALE

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ABSTRACT Dandy-Walker Malformation (DWM) is a rare congenital malformation characterized by hypoplasia of the cerebellar vermis and its upward rotation and cystic enlargement of the fourth ventricle. The clinical manifestations include headache, psychomotor retardation, ataxia, urinary incontinence and hydrocephalus. This is a case of 25 years male presented to general medicine opd in our institute with intermittent episodes of nausea, vomiting, seizures, ataxia for last few years which was further evaluated by CT and MRI of brain in Dept of Radiodiagnosis suggesting diagnosis of Dandy-Walker malformation. patient was referred to Dept of Neurology for exact management.

KEYWORDS : Dandy-Walker Malformation, Dandy-Walker Variant, Cerebellar Vermis Hypoplasia.

INTRODUCTION:

Dandy–Walker malformation is a rare congenital abnormality of the posterior cranial fossa and the incidence of Dandy–Walker syndrome (DWS) is 1:25,000–1:35,000 live births.[1] .The disorder was originally described in 1887 by Sutton. Later, it was explained by W. Dandy and K. Blackfan in 1914 followed by Tagart and Walker in 1942, and finally, C. Benda in 1954 designated this disorder as DWS.[2,3] DWM may be asymptomatic or associated with various diseases such as bipolar disorder, Acquired Immunodeficiency Syndrome, and kidney and liver diseases.4 Anaesthetic management of DWM patients may be faced with severe challenges due to multi-organ association of craniofacial abnormalities, hydrocephalus, renal, and cardiac anomalies.5

Case Report:

A 25 years male was referred to Department of Radiodiagnosis of our institute for evaluation of intermittent episodes of nausea, vomiting, ataxia and seizures . He was the second child of her family and did not have any significant family history of congenital disorders, additionally, her mother did not remember any exposure to drugs or infections during her pregnancy.

Initially Brain computed tomography (CT) scan, which had been done, showed cystic dilatation of the fourth ventricle with enlargement of the posterior fossa, evidence of hypoplastic cerebellar vermis (Figure 1). Then the patient underwent magnetic resonance imaging (MRI) of brain and it showed cystic dilatation of the fourth ventricle with enlargement of the posterior fossa (Figure 2A), evidence of hypoplastic cerebellar vermis with cephalad rotation of the vermian remnant (Figure 2B), vermian hyplasia , large posterior fossa communicating with fourth ventricle (Figure 2C).These findings confirmed the diagnosis of DWM.

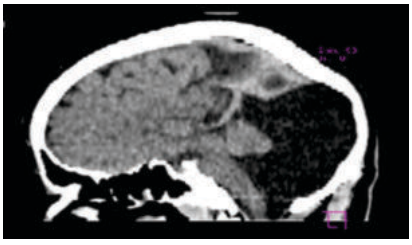

Figure-1: NCCT Brain show large posterior fossa cyst communicated with 4th ventricle, cerebellar vermis hypoplasia, tentorial apex and sinus confluence are elevated.

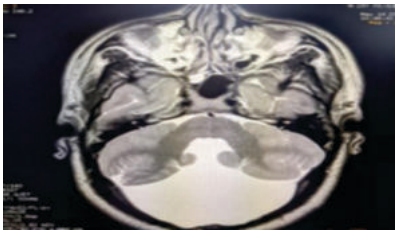

Figure 2A: Axial T2 MR showing cystic dilatation of fourth ventricle.

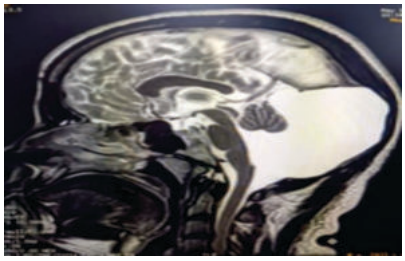

Figure 2B: Sagittal T2 MR ,Evidence of hypoplastic cerebellar vermis and cephalad rotation of vermian remnant.

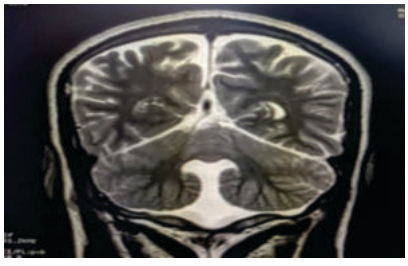

Figure 2C: Coronal T2 MR Showing vermian hyplasia , large posterior fossa communicating with fourth ventricle.

DISCUSSION:

Dandy-Walker syndrome is a rare malformation defined as dilatation of the posterior fossa, cystic enlargement of the fourth ventricle, hypoplasia of the cerebellar vermis and its upward displacement. DWS is normally presented in childhood, however, rare cases are also reported in adulthood.6 Predisposing factors of DWS are common congenital infections such as toxoplasmosis, cytomegalovirus and rubella, and drugs like, warfarin, retinol derivative (isotretinoin) and ethanol.[7,8] However, we are not certain whether the mother of this patient had faced these factors or

not.Our case had recurrent nausea, vomiting, headache and seizure, in addition mental retardation and macrocephaly. About 90% of individuals with DWM have hydrocephalus, 15.30% have seizure, 41.71% have poor intelligence development, increased head circumference, ataxia, muscle rigidity, and growth retardation at the time of diagnosis.7 The diagnosis of DWM is made by computed tomography and MRI.9 We observed cystic enlargement of the posterior fossa and a dilated fourth ventricle in MRI images of the patient.

Differential diagnosis:

Dandy-walker variants:

Posterior fossa volume will be normal.

Mega cisterna magna:

Vermis,fourth ventricle and Posterior fossa volume will be normal with normal tegmento-vermian angle.

Arachnoid Cyst:

No communication with fourth ventricle.

Blake pouch cyst:

Vermis will be normal. Hydrocephalus may be present.

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