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Dandy-Walker Variant: A Case Report

Dr. Sreelatha S[1], Dr. Vedavathy Nayak[2], Dr. Sathya P[3], Dr. Nirmala Hanji[3]
[1]Associate Professor, Dept of OBG, ESICMC & PGIMSR, Rajajinagar, Bangalore, India
[2]Assistant Professor, Dept of OBG, ESICMC & PGIMSR, Rajajinagar, Bangalore, India
[3]Junior Resident, Dept of OBG, ESICMC & PGIMSR, Rajajinagar, Bangalore, India

***Corresponding Author:**
Name: Dr. Sreelatha.S
Email: dr.sreelatha2011@gmail.com

Abstract: Dandy–Walker malformation is a rare abnormality of the central nervous system (CNS) with a reported incidence of 1 in 30000 live births and a slight female predominance. Dandy Walker variant forms part of the spectrum of Dandy Walker malformation. It is characterized by partial agenesis of the vermis along with mild expansion of the foramen magnum.
Keywords: Dandy–Walker, malformation, cerebellar vermis.

INTRODUCTION

The term Dandy-Walker syndrome or malformation was originally described in 1914. Dandy Walker malformation represents a rare developmental abnormality of the rhombencephalon with complete or partial agenesis of the cerebellar vermis, and cystic dilatation of the fourth ventricle and the posterior cranial fossa [1]. Coexisting structural and chromosomal abnormalities occur frequently and adversely affect survival.

CASE REPORT

A 35 year old lady G2P1L1 presented to the antenatal OPD at 20 weeks of gestation. She was married for the last 6 years with no h/o consanguinity. Her first pregnancy and delivery were uneventful and she had a 4 year old daughter. A routine antenatal scan at 20 weeks showed the following features: SLIUG of 20weeks 5days growth with

- Cephalocele in occipital region
- Posterior fossa cyst in the fetal brain
- Monoventricle of the fetal heart with truncus arteriosus
- Hypoplastic mandible and nasal bone

The diagnosis was Dandy-Walker Syndrome Variant.

The patient opted for termination of the pregnancy in view of the anomalies. The patient was not willing for post-mortem and karyotyping of the foetus.

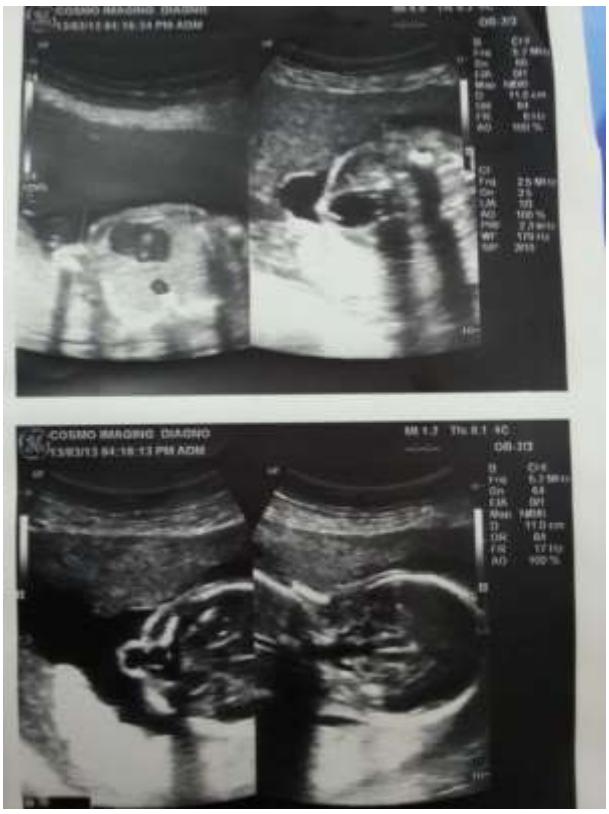

Fig-1: Dandy-Walker malformation

DISCUSSION

Dandy-Walker malformation was first described in 1914. Dandy-Walker malformation is characterised by failure of development of the cerebellar vermis with resulting communication between the fourth ventricle and the cisterna magna, with an associated midline cyst and cerebellar hypoplasia. Dandy-Walker malformation is exceedingly rare, with an estimated incidence of about 1:30000

births, and is found in 4-12% of all cases of infantile hydrocephalus [2]. However, minor variations of this condition are frequently encountered which include Dandy-Walker variant and mega cisterna magna. Dandy-Walker malformation, Dandy- Walker variant and mega cisterna magna are all steps along a continuum of abnormality of the posterior fossa and these conditions are grouped under the umbrella term, Dandy-Walker complex [3].

Dandy-Walker variant which is characterised by partial agenesis/hypoplasia of the vermis and mild expansion of the foramen magnum is more common. Ventriculomegaly is found in approximately one –third of cases.

Dandy-Walker complex is frequently associated with neural tube defects such as agenesis of the corpus callosum and holoprosencephaly. Other deformities include encephaloceles, polycystic kidneys, and cardiovascular defects. Chromosomal abnormalities in particular, trisomies 13, 18, 21 are frequently associated [4].

The prognosis for Dandy-Walker malformation or variant is generally poor. The main features which impact prognosis are karyotype and the presence of additional abnormalities on scan. In a study of 47 foetuses diagnosed with Dandy-Walker malformation or Dandy-Walker variant, 41 (82%) had associated anomalies, 44 died (94% mortality rate) and all 3 survivors had serious neurodevelopmental handicap [5]. It is also known that foetuses with a Dandy-Walker malformation diagnosed prenatally have higher morbidity and mortality rates than children with postnatal diagnosis of the same defect [6]. As a consequence it seems likely that early prenatal recognition of Dandy- Walker malformation would be associated with a worse prognosis than late prenatal diagnosis.

The accurate diagnosis of antenatal conditions is essential not only for the management of the current pregnancy but also for future pregnancies to aid in the identification of inheritable diseases and provide information for accurate pre-natal counselling.

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