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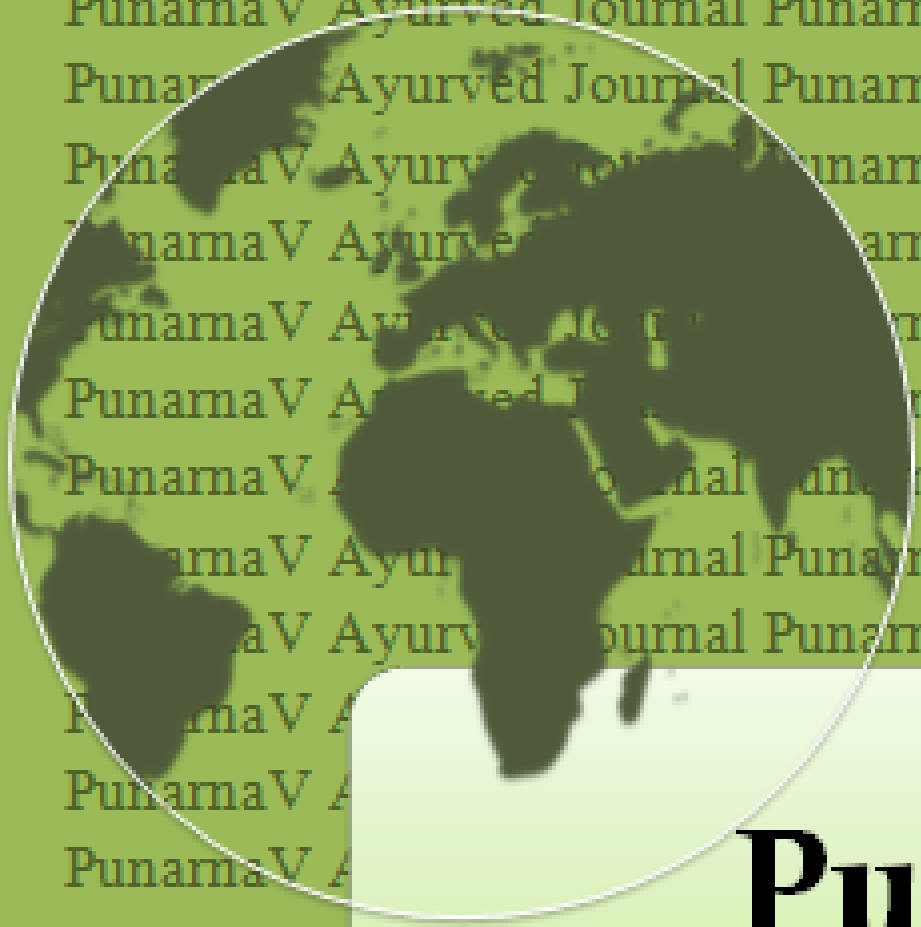

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DANDY WALKER MALFORMATION: A CASE REPORT

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ABSTRACT:

Dandy–Walker syndrome is a congenital rare disorder characterized by complete or partial agenesis of the vermis, cystic dilatation of the fourth ventricle and an enlarged posterior fossa1. The precise etiology is unknown, although there have been reports of associations with risk factors like maternal virus infections (rubella, toxoplasma, and cytomegalovirus) and alcohol consumption. The reported incidence varies between one per 2500 births to one per 100,000 births. Although there is an extensive list of signs attributed to DWM, final diagnosis is solely dependent on imaging techniques as there are no signs that are characteristic of DWM. This article reports a case with DWM who was diagnosed by ultra sound sonography.

Key words: Dandy walker syndrome, vermis, fourth ventricle.

INTRODUCTION

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Obstetrics is the art and science of caring for women and their unborn progeny during pregnancy, labour, immediate puerperium etc. Approximately 2% of new born infants have a major congenital malformation, making this the leading cause of infant mortality. Survivors are usually afflicted by lifelong disabilities, resulting in the need for frequent hospital admissions and long term care. Advances in ultrasound technology have made possible the prenatal identification of an increasingly large number of fetal congenital malformations. The current method for the detection of fetal congenital abnormalities is a comprehensive ultrasound examination or fetal anatomical survey performed between 18 and 22 weeks of gestation[2].

An adequate ultrasound examination of fetal central nervous system requires visualization of the cerebellum and the cisterna magna, the lateral ventricles,the choroid plexuses the thalami, third ventricle,cavum septum pellucidum and the falxcerebri. Most of this information can be obtained with a transverse scan at the level of septum pellucidum, a coronal view perpendicular to the transverse scan and a suboccpito bregmatic view. The CNS abnormalities most frequently found during the 18-22 weeks sonographic examination are anencephaly, spina bifida, ventriculomegaly, holoprosencephaly, Dandy –walker malformation etc.

In this present article a case has been reported which was diagnosed as Dandy – Walker malformation with a detailed discussion on this topic.

CASE REPORT

A moderately built female of age 24 years, with a marital history of 6 years, residing at Hassan , c/o amenorrhea since 6 months, was under regular ANC check up, approached our OPD on 05/01/2015 for her booked ANC visit. Her LMP was on 12/07/2014 and based on that EDD will be on 19/04/2015. Her previous MH was regular (before conception).

Her obstetrical history reveals G4 P1 L1 A2 D0. In her first pregnancy she had a full term normal delivery of a female baby, now at the age of 4 years. In her second pregnancy she underwent medical

termination of pregnancy at the second month (3 years back). In her third pregnancy she had missed abortion at 1 ½ month of gestation (3 months back). And the fourth one is the present pregnancy.

From first month onwards she was under regular ANC visit. She was suggested for anomaly scam on 05/01/2015.It suggested the impression as single live fetus of 22 weeks 4 days seen in cephalic presentation, with fetal cranium was presented with hydrocephalus. Spine / lumbar region shows spins bifida, with meningiocoel. As per ultrasound scan report; the case was diagnosed as Dandy-Walker malformation.

While analyzing her previous scan report on 08/10/2014 it suggested normal study with NT : 2.2 mm, with single live intrauterine gestation of 11 weeks 6 days +/- 1 week. As per the management protocol, she was suggested for termination of pregnancy.

DISCUSSION

Dandy–Walker malformation was originally described in 1887 by Sutton and further characterized by Dandy and Black fan in 1914 followed by Tagart and Walker in 1942. Benda finally labeled this disease as dandy walker in 1954.

Total cerebellar agenesis is a rare malformation, which may be due to defective early patterning of the hind brain. In contrast, hypoplasia or aplasia of the cerebellar vermis is relatively common and is associated with cystic dilatation of the fourth ventricle[3].This abnormality is due to incomplete fusion of the cerebellar anlage in dorsal midline. Dandy –Walker malformation is frequently associated with other abnormalities of the CNS. It can be an isolated finding or it may be associated with chromosomal abnormalities, single – gene disorders[4] or congenital infections(e.g. Cytomegalovirus, toxoplasmosis, rubella) and exposure to alcohol.

The syndrome includes three classical features: hydrocephalus, posterior fossa cyst and defect in cerebellar vermis. It accounts for approximately 5-10% of case of hydrocephalus. Approximately 80% of patients with Dandy –Walker malformation present with ventriculomegaly. However the degree of ventricular dilatation varies and same cases with open foramina of Lushka and Magendie may never develop ventricular dilatation in utero. The malformation should be suspected when there is marked increase in the diameter of cisterna magna (more than 10 mm from the cerebellar vermis to the inner border of skull) and an absence of cerebellar vermis with or without associated ventricular dilatation of

the cisterna magna as seen in cases of trisomy 18[5].

The incidence of associated cerebral defects is as high as 68%. A common associated defect is the absence of the corpus callosum which is evidenced by an inability to visualize the cavum septum pellucidum. Chromosomal abnormalities are present in approximately 20% of cases and include trisomies 13, 18 and 21.

The prognosis of foetus diagnosed with Dandy –walker malformation is poor. Approximately 30% of them will die in utero or in the immediate neonatal period. The outcome of survivors depends on the presence of associated anatomic or chromosomal abnormalities. Infants with DWM may present with early signs such as vomiting, sleepiness, irritability, convulsions, unsteadiness and lack of muscle coordination. The clinical manifestations include psychomotor and growth retardation, hypotonia, strabismus, myopia, a short neck, microcephaly, brachycephaly, hypertelorism, antimongoloid slant of palpebral fissures, globulus large nose, large mouth with down turned corners, poorly lobulated ears, high arch palate, cleft palate, small hands and feet, clinodactyly, and the brachymesophalangy of the little fingers. When the defect is isolated with no hydrocephaly, expectancy is the usual treatment, otherwise protocol suggested was termination of pregnancy.

CONCLUSION

Dandy–Walker syndrome is a rare yet serious health issue which attributes its effect on the future of mother and child.. Many aspects of this malformation remain poorly understood. More research studies are required in order to explore the demographic and environmental factors that might influence different aspects of this anomaly. Even though it is rare the important finding in the USG warrants a detailed search to elucidate the reasons, probably in a larger study with a prospective approach.

REFERENCES

1.Jyothi.T ,Santhoshkumar, Mamatha.G.P.Dandy-Walker malformation.Indian Journal of Human Genetics.2010 Jan-Apr;16(1):33-35.

2. Fernando Arias .Practical Guide to High Risk Pregnancy and Delivery, A South Asian Prespective .thirdedition.India:Reed Elsevier India Private Limited.2008;62-63.

3.Geoffery Chamberlain, Philip.J.Turnbull'sObstertrics. Third edition.China:Churchill Livingstone.2001;54-55.

4.Geoffery Chamberlain, Philip.J.Turnbull'sObstertrics. Third edition.China:Churchill Livingstone.2001;175-176.

5.Fernando Arias .Practical Guide to High Risk Pregnancy and Delivery, A South Asian Prespective .thirdedition.India:Reed Elsevier India Private Limited.2008;69.