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AQUEDUCTAL STENOSIS- A RARE CASE REPORT

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ABSTRACT Aqueductal stenosis is a narrowing of aqueduct of sylvius which block the CSF in the ventricular system. It is the narrowest part of the CSF pathway. Blockage of aqueduct can lead to hydrocephalus. A 25-year-old patient, Gravida 2 Para 1 Living 1 with history of 5 months of amenorrhea came to OBG Outpatient department for antenatal checkup for the first time. Anomaly scan showed A single live intrauterine fetus of 23 weeks period of gestation with grossly dilated lateral and third ventricle suggestive of aqueductal stenosis. Patient was counselled regarding the congenital anomaly and termination of the pregnancy was done. Aqueductal stenosis with hydrocephalus contributes a significant burden of morbidity in patients. All pregnant women are advised for regular antenatal check-up and targeted anomaly scan between 18-24 weeks is advised.

KEYWORDS : Aqueductal stenosis, Anomaly scan, Hydrocephalus

INTRODUCTION

Aqueductal stenosis is a narrowing of aqueductus of sylvius which block the CSF in the ventricular system. The aqueduct of sylvius connects the third ventricle to fourth ventricle. It is the narrowest part of the CSF pathway. Blockage of aqueduct results in accumulation of CSF which leads to hydrocephalus. Hydrocephalus resulting from obstruction within the ventricular system is known as Obstructive or Non-communicating hydrocephalus.

Aqueducatal stenosis is a rare condition and the incidence is 1 in 5000 babies. In most cases it is sporadic in occurrence and in others genetic and viral infections were found as a causative factor. 10% of males with aqueductal stenosis had genetic cause as it is usually linked with to X chromosome. If a genetic factor is the cause the chance of occurance in next pregnancy is 25-50%.

CASE REPORT

An unbooked case of 25 year old patient, resident of Raichur, Gravida 2 Para 1 Living 1 with history of 5 months of amenorrhea came to OBG Outpatient department for antenatal checkup for the first time.

General physical examination and systemic examination was uneventful.

Per abdomen examination-Uterus corresponds to 20-22 weeks period of gestation, relaxed, FHS heard on auscultation, external ballottement present.

Investigations- Hb-11.4g%, TLC-6200, PC-2.13 lakh, Blood grouping and typing -A positive, Urine routine- normal, Serology- non reactive, BT/CT- normal, TSH-2.11uIU/L.

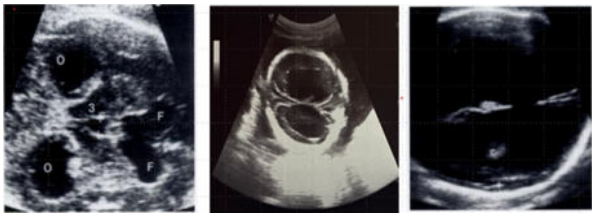

Figure-1: A) Fetal brain showing Dilatation of both lateral and third ventricles with aqueductal stenosis F-frontal horn; O-occipital horn; 3-third ventricle. B) Fetal brain showing Dilated lateral ventricles. C) Fetal brain showing marked hydrocephalus with cortical thinning C-cortical thinning

Anomaly scan showed a single live intrauterine fetus of 23 week period of gestation with grossly dilated lateral(20mm) and third ventricle with dangling choroids and fenestrated falx suggestive of aqueductal stenosis. Mildly hypoplastic nasal bone. Tiny septal defect of interventricular septum seen superior to semilunar valve-suggestive of oulet VSD. Persistant left superior venacava. Placenta-fundal posterior grade 2, AFI-adequate, Fhr-140bpm, fetal weight of 473 gm.

MANAGEMENT

Patient was counselled regarding the congenital anomaly, prognosis of the condition and termination of the pregnancy was advised

Pregnancy was terminated with tablet Mifepristone 200mg on day-1. Foley's induction was done on day 2.She expelled a single dead female fetus of birth weight 550gm with placenta intoto.

On gross examination the baby had hydrocephalus. Fetal autopsy could not be conducted due to patient refusal

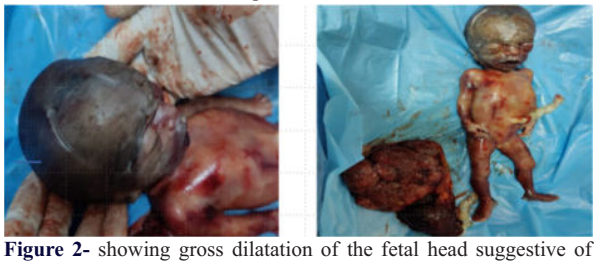

Figure 2- showing gross dilatation of the fetal head suggestive of hydrocephalus

DISCUSSION

Congenital aqueductal stenosis is one of the cause of prenatal obstructive hydrocephalus. The etiology is multifactorial including both genetic and acquired forms. Acquired cause can be intrinsic (obstruction of aqueduct lumen) or extrinsic (external compression).

Prenatally acquired causes are mostly intrinsic resulting from infection (aqueductal gliosis/web) or intraventricular haemorrhage. Extrinsic causes are less common prenatal period and include tectal plate mass, periaqueductal vascular malformation or compression from ventricular diverticulum.

Of genetic cause, X linked mutation in the gene for L1 cell adhesion molecule is known to be associated with CAS. Most commonly seen in Male fetus.

They are at an elevated risk of long term neurological dysfunction. Neurological deficits in Congenital aqueductal stenosis result mainly due to CSF obstruction and ventriculomegaly.

Long term sequelae like motor abnormalities, disabilities, epilepsy, endocrine dysfunction (delayed puberty, amenorrhea) can be seen.

There is little progress on antenatal management of ventriculomegaly like ventriculo-amniotic shunt. Postnatally the modalities of

intervention for aqueductal stenosis include ventriculoperitoneal shunting and endoscopic third ventriculostomy

CONCLUSION

Aqueductal stenosis with hydrocephalus contributes to a significant burden of morbidity in patients due to its long term neurological and endocrine sequelae. Hence all pregnant women are advised for regular antenatal check-up and get NT scan at 11-13+6 weeks and targeted anomaly scan between 18-24 weeks.

Early diagnosis of aqueductal stenosis with hydrocephalus can help in making decision about termination of the pregnancy as the prognosis is poor with mortality rate of 40% and two-third of the surviving children have variable mental and motar diasabilities even with appropriate treatment

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