International Journal of Science and Research (IJSR)
ISSN: 2319-7064
SJIF (2022): 7.942

Case Report of Arnold Chiari Malformation

Dr. Chakka Yamini Uma Deepthi[1], Dr. Sai Charitha[2]

[1]Junior Resident, Sree Balaji Medical College and Hospital, Chennai, India
Corresponding author Email id: *visakshi274521[at]gmail.com*
Ph no - 9440027812

[2]Senior Resident, Sree Balaji Medical College and Hospital, Chennai, India

Abstract: ***The Arnold Chiari malformation a congenital abnormality of Central nervous system, characterized by the downward displacement of parts of the cerebellum, fourth ventricle, pons and medulla oblongata into the spinal canal. This malformation is one of the causative factor of death in neonates and infants. The malformation is named after Austrian pathologist Hans Chiari and German pathologist Julius Arnold, as Arnold–Chiari malformation. In this case report, we present A Chiari malformation II detected at 19 weeks of gestation by routinely sonographic screening. The role of antenatal ultrasonography in identifying the malformation are discussed.***

Keywords: Arnold Chiari malformation, congenital abnormality, central nervous system, banana sign, Lemon sign

1. Introduction

Occurs more in female baby's than in male.

More prevalence in certain group of people including Celtic descent (Britain, Ireland, Wales, Scotland, Cornwall, Isle of Man). Arnold - Chiari malformation, which has an incidence of 0.5 per 1000 livebirths, is one of the causes of CNS abnormalities, accounting for 4% of all abortions and a 1 - 4% recurrent risk, and is classified into three types.

Type I consists inferior displacement of tonsils and cerebellum without displacement of the fourth ventricle or medulla.

Arnold Chiari malformation type II is more common in neonates and infants and is defined by the displacement of cerebellar tonsils, cerebellar fourth ventricle, pons, and medulla oblongata through the foramen of magnum into the spinal cord cannal. This is commonly linked to hydrocephalus and myelomeningocele.

Chiari III malformation has a high cervical encephalomeningocele that includes the medulla, fourth ventricles, and nearly the entire cerebellum. Frontal bone scalloping (lemon sing) and absence or abnormal anterior curvature of the cerebellar hemispheres are the most obvious usg findings (banana sign) A 22 years old pregnant woman, primigravida, spontaneous conception with no history of the 0familial genetic disorders was admitted to Sree Balaji Medical College and Hospital (Chrompet, Chennai). First ANC visit was at 7 weeks & patient was advised folic acid supplementation and Nuchal scan. between 7 weeks to 19 weeks patient had no work up due to irregular follow up. In ultrasonographic examination at 19 weeks of gestation, multiple fetal anomalies include lemon sign, banana sign, mild ventriculomegaly, with open lumbar myelomeningcoele (2.07 X 1.16cm) were detected. According to these ultrasonographic findings, the Arnold Chiari malformation (type II) was confirmed and termination of pregnancy performed at 19 weeks

2. Discussion

Multiple studies have evaluated the accuracy of sonography of diagnosis of Chiari malformations.

Routine usg is performed in the second trimester has a detection rate of approximately 60% for fetal congenital abnormalities

The CNS abnormalities are one of the most commonly detected. Chiari malformation is one among the CNS abnormalities diagnosed with antenatal ultrasonography.

The frontal bone scalloping (lemon sign) and absence or abnormal curvature of the cerebellar hemispheres are ultrasonographic findings (banana sign)

Characteristic brain findings (lemon sign and banana sign) can be seen on antenatal ultrasonography as early as 12 weeks, and myelomeninogocoele can be identified as early as 10 weeks. Downward traction of the brain caused by compression from the third and fourth ventricles results in a reduction in the anterior calvarium, resulting in a triangular shaped head in the biparietal diameter, similar to LEMON. .

CSF leak and low intracranial pressure. This leads to effacement of the posterior fossa and cerebellum accommodates to the small space. The cerebellum wraps around the brain stem and appears as "BANANA

The diagnosis of myelomeningocele in a fetus is important since it provides the parent an opportunity to consider pregnancy termination. Among parents electing to continue the pregnancy, adequate counselling and psychological preparation should be provided. Hydrocephalus and tethered cord determine the prognosis for deterioration.

3. Conclusion

The ultrasonographic prenatal screening is selected as the main primary method of assessment of the early fetal malformations. Early diagnosis of such malformations helps

to make decision to offer further fetal karyotyping or termination of pregnancy.

Arnold Chiari malformation that can be prevented by preconceptional folic acid and vitamin B12 supplementation. In a developing country like India pregnancies are unplanned. At 6 weeks of gestation, the rostral and neural pores close. Folic acid supplementation that is delayed will miss a critical period of organogenesis, neural tube closure, and thus the defect. In view of recurrence of 1 to 2 %, genetic counselling and preconception folic acid, 1st trimester screening and proper follow up of the pregnancy should be done so as to plan proper management of the pregnancy.

References

[1] Woodward P, Kennedy A, Sohaey R, Byrne JLB, Oh KY, Puchalski MD. Diagnostic imaging.1sted. Canada, Amirsys Elsevier Saunders.2005; chapter 2: 21
[2] Silva SR, Jeanty P. In: Callen PW, ed. Ultrasonography in obstetrics and gynecology. eds. Gynecologic, obstetric and
[3] breast radiology. A text/Atlas of imaging in women.4th ed. Blakwell science, Oxford, 2000; 73
[4] Ganesh D, Sagayaraj BM, Barua RK, Sharma N, Ranga U. Arnold Chiari Malformation with Spina Bifida: A Lost Oppurtunity of Folic Acid Supplementation. J Clin Diagn Res.2014 Dec; 8 (12): OD01 - 3. Doi: 10.7860/JCDR/2014/11242.5335. Epub 2014 Dec 5
[5] Nyberg DA, Souter VL. Sonographic markers of fetal trisomies: second trimester. J Ultrasound Med.2001 Jun; 20 (6): 655 - 74 (PubMed)
[6] Irureragoyena JI, Trampe B, Shah D. Prenatal diagnosis of chiari malformation with syringomyelia in the second trimester.
[7] J Matern Fetal Neonatal Med.2010 Feb; 23 (2): 184 - 6. doi: 10.3109/14767050903061769.