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Arnold Chiari Malformation: A Case Report

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Abstract
The Arnold Chiari malformation is a congenital abnormality of Central nervous system, characterized by downward displacement of the parts of the cerebellum, fourth ventricle, pons and medulla oblongata into the spinal canal. 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 18 weeks of gestation by USG screening. The role of prenatal sonography in recognition of malformation are discussed.

Introduction

Occurs more often in female baby than in male.

More prevalent in certain groups including people of Celtic descent (Britain, Ireland, Wales, Scotland, Cornwall, Isle of man).

It has an incidence of 0.4 in 1000 live births and is one of the CNS abnormalities that is responsible for 3 % of all abortions and 1 – 2 % recurrence risk(1). Chiari classified the hindbrain malformations into type I,II , III and IV(2). Chiari II is usually associated with hydrocephalus and myelomeningocoele(3).

The obvious sonographic findings are frontal bone scalloping (lemon sign) and absent cerebellum or abnormal anterior curvature of the cerebellar hemispheres (banana sign)(4).

Case Report

A 23 years old pregnant woman, primigravida, spontaneous conception with no history of familial genetic disorders was admitted to MOOLCHAND MEDCITY (lajpat nagar, new Delhi). First ANC visit was at 8 weeks & patient was advised folic acid supplementation and Nuchal scan. between 8 weeks to 18 weeks patient had no work up due to her individual problem. In sonographic monitoring at 18 weeks of gestation, multiple fetal anomalies include *lemon sign (fig 3), banana sign* (*fig 4*),mild *ventriculomegaly*, with *open lumbar myelomeningcoele (2.07 X 1.16cm)(fig 1 (a)& (b))* were detected. According to these sonographic findings, the Arnold Chiari malformation (type II) was confirmed and termination of pregnancy performed at 18 weeks

of gestation and a live male fetus was delivered vaginally of 220gm (fig 2)

Fig 1 (a) : Lumbar Myelomeningocoele

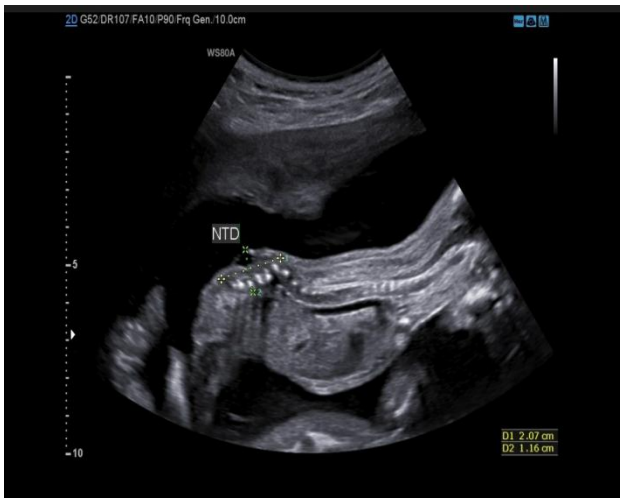


Fig 1 (b)

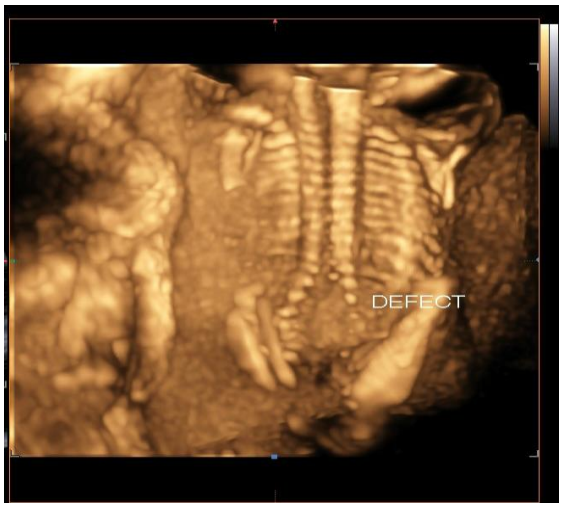
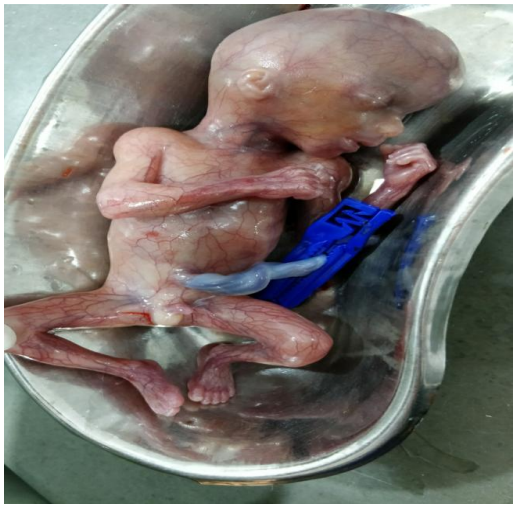
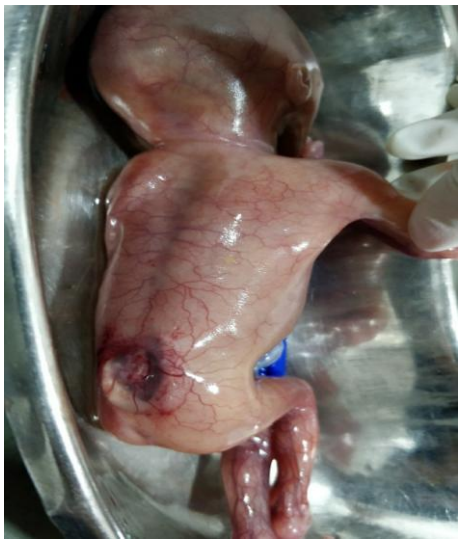


Fig 2 : Live Male Fetus





Discussion

Multiple studies have evaluated the accuracy of sonography of diagnosis of Chiari malformations [5-7]. Routine ultrasound performed in the second trimester has a detection rate of approximately 70 – 90% for fetal congenital abnormalities[8]. The CNS abnormalities are one of the most common ones detected. Chiari malformation is one among the CNS abnormalities diagnosed with prenatal sonography[9]. The sonographic findings are the frontal bone scalloping (lemon sign) (Fig 3) and absent cerebellum or abnormal curvature of the cerebellar hemispheres (banana sign) (Fig 4)[10] . On prenatal sonography, characteristic brain findings (lemon sign and banana sign) may be seen as early as 12 weeks and myelomeningocoele may be identified as early as 10 weeks.

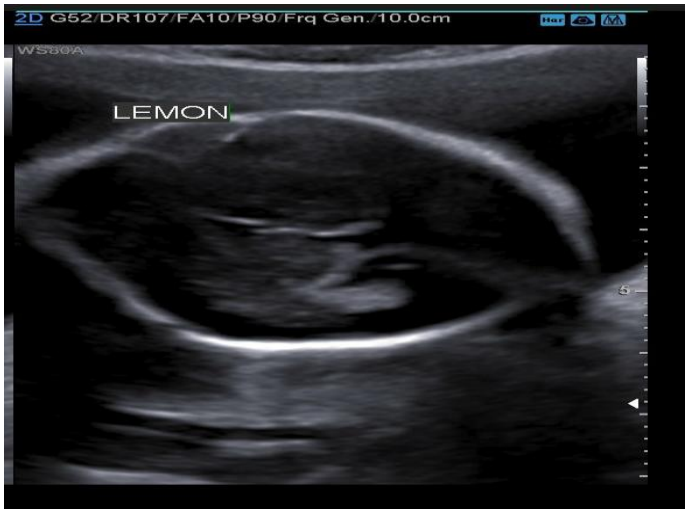


Fig 3 – Lemon Sign: Herniation of cerebellar tonsils and midbrain structures into the foramen magnum,causing ventriculomegaly due to

compression from 3rd and 4th ventricles, downward traction of the brain causes a reduction in the anterior calvarium, resulting in a triangular shaped head in the biparietal diameter like a "LEMON

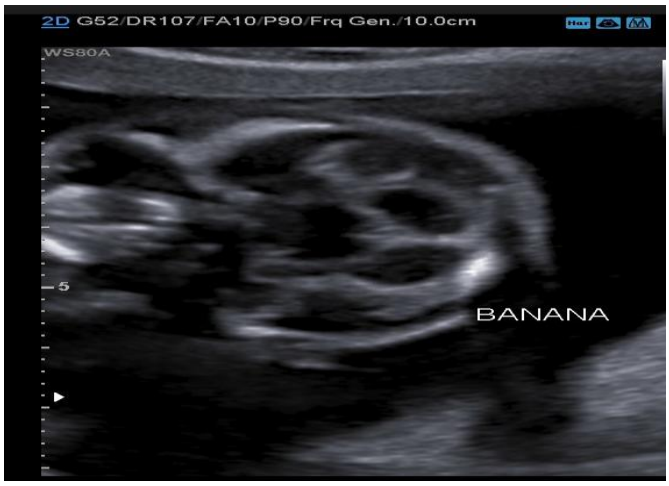


Fig 4 – Banana Sign: Spinal dysraphism leads to CSF leak and low intracranial pressure. This leads to effacement of the posterior fossa and cerebellum accommodates to the small space. The cerebellum wraps around the brain stem and appears as "BANANA

There are 4 types of Arnold chiari malformation – types I - IV[(11)]

Type I – Herniation of only cerebellar tonsil, not assosciated with myelomeningocoele

Type II – Herniation of cerebellar vermis and brain stem into the spinal canal with spina bifida and is always associated with neural tube defects like myelomeningocoele.

Type III – Rare type of brain herniation assosciated with cephalocoele or cranio cervical meningocoele

Type IV– Extreme cerebellar hypoplasia and caudal displacement of posterior cranial fossa contents[(11)].

In view of severe morbidity assosciated with these malformations, a daily intake of folic acid is recommended in the preconception period. A large meta analysis study strongly suggests that polymorphism of genes involved in folate metabolite pathway is strongly related to neural tube defects[(12)]. Many theories have been proposed regarding etiology and causative associations between descending cerebellar herniations and myelomeningocoele, however none of them could conclude in detail about origin and exact mechanism of assosciation[(13)].

The diagnosis of myelomeningocoele 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[(14)]. Hydrocephalus and tethered cord determine the prognosis for deterioration.

Conclusion

The ultrasonographic prenatal screening is emphasized as the primary method of assessment of the early fetal malformation. 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[(15)]. In India most of the pregnancies are unplanned. The rostral and neural pores close at 6 weeks of gestation. A delayed folic acid supplementation is bound to miss the vital period of organogenesis and neural tube closure, and hence the defect. Recurrence of 1 to 2 % should be remembered also for the same lady who suffered so that folic acid, ultrasound, planned pregnancy can be done in time.

Concluding, early ultrasonography in planned pregnancy with early folic acid and B12 supplementation is suggested to prevent central nervous system anomalies like Arnold Chiari malformation.

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