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Case Study

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A CASE REPORT OF ULTRASOUND-BASED DIAGNOSIS OF ARNOLD CHIARI TYPE MALFORMATION IN A SECONDARY REFERRAL CARE HOSPITAL

Vivek Kumar Garg[1] and Manjula Sharma[2]*

[1]Department of Radiodiagnosis, NSCB Zonal Hospital Mandi, Himachal Pradesh, India.
[2]Medical Officer, Civil Hospital, Sundernagar, Himachal Pradesh, India.

***Corresponding Author: Manjula Sharma**

Medical Officer, Civil Hospital, Sundernagar, Himachal Pradesh, India.

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ABSTRACT

Arnold Chiari malformation is a rare congenital anomaly of central nervous system characterised by caudal displacement of brainstem and cerebellum. Arnold Chiari malformation is one of the common cause of congenital hydrocephalus. It is pertinent to have thorough understanding of sonographic features for diagnosis of Arnold Chiari malformation in a developing foetus. In this case report, we present a Arnold Chiari III malformation detected at 19 weeks with routine sonographic examination. Role of detailed pre-natal ultrasound scan in diagnosing Arnold Chiari malformations is discussed in this case report.

KEYWORDS: Arnold Chiari malformation III, prenatal diagnosis, Ultrasonographic screening.

INTRODUCTION

Arnold Chiari malformation was first discovered by Hans Chiari in 1891in a paediatric specimen.[1] The risk for this malformation is 0.4:1000 live births leading to 3% of all abortions with 1-2% of recurring risk. Arnold Chiari malformation was initially classified into three types with rest of the types added later on.[2] Chiari I malformation is the most common, consisting of peg-like cerebellar tonsils displaced into the upper cervical canal through the foramen magnum. Chiari 1.5 malformation consists of caudal descent of cerebellar tonsils and brainstem. Chiari II malformation comprises displacement of medulla, fourth ventricle and cerebellar vermis through foramen magnum and is usually associated with lumbosacral myelomeningocele. Chiari IV malformation represents severe cerebellar hypoplasia without caudal regression of cerebellum. Chiari V malformation consists of absent cerebellum with herniation of occipital lobe through foramen magnum while Chiari 0 represents syrinx without cerebellar or brain stem descent.[3] Chiari III malformation represents malformations similar to type II with an occipital and /or high cervical encephalocele. We report a case of Type III Chiari malformation detected at 19 weeks of gestation in routine antenatal ultrasound scan at NSCB Zonal Hospital Mandi, Himachal Pradesh, India.

CASE REPORT

A 22 year-old, pregnant women, primi-gravida with no history of familial genetic disorders underwent routine antenatal check-up at 22weeks at NSCB Zonal Hospital Mandi (Department of Gynaecology). She was referred to Department of Radiodiagnosis for routine level II screening. Previous sonographic screening at 13 weeks confirmed normal foetal anatomy. In routine sonographic monitoring at 19 weeks, multiple foetal anomalies were noted including microcephaly, lemon sign occipital/high cervical encephalocele(figure 1), mild to moderate ventriculomegaly, , herniation of posterior fossa contents and club foot. The patient had concurrent α-Fetoprotein levels done, showing increased values(180mmol). In accordance with these sonographic/biochemical findings Chiari type III malformation was confirmed and termination of pregnancy was performed at 19weeks4days.

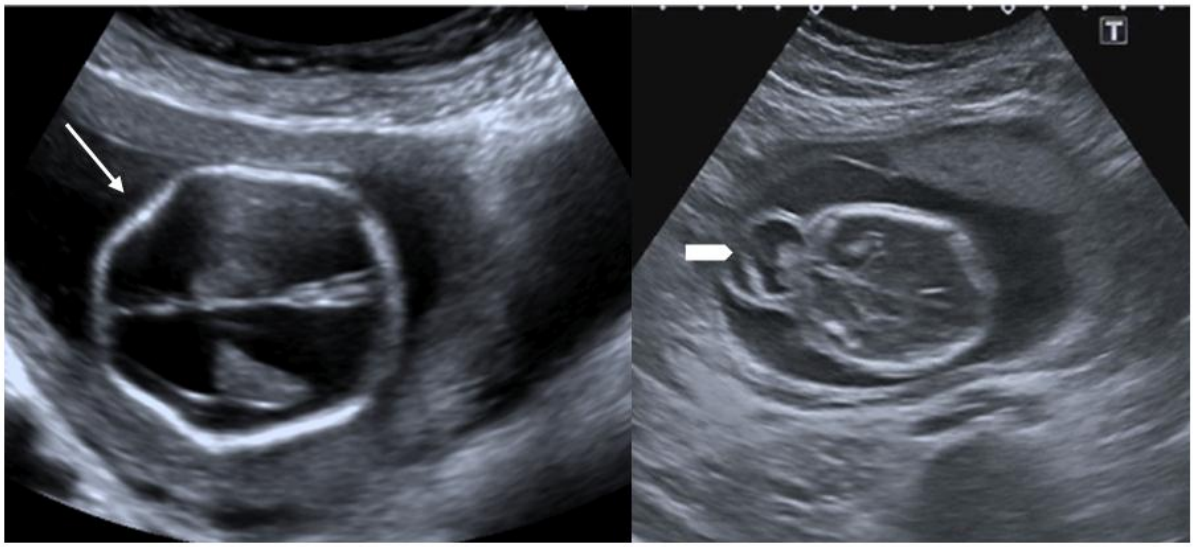

Fig1: The lemon sign(arrow) and occipital meningocele(arrowhead)

DISCUSSION

There are multiple studies mentioned in literature about accuracy of ultrasound in the diagnosis of various Chiari malformations.[4,5,6]

The features of Chiari III are combinations of Chiari II with occipital/high cervical encephalocele. Chiari II malformation is basically a disorder of neural tube closure. Most characteristic findings of Chiari II are of infratentorial origin including small and overcrowded posterior fossa(PF) with effacement of cisterna magna[5] with deformation of cerebellum known as " Banana sign". With an overcrowded posterior fossa, fourth ventricle appears effaced leading to ventriculomegaly. A "cascade of tissue" is usually seen herniating through foramen magnum with a medullary spur. The cerebellar hemispheres and vermis are pushed upward through incisura leading to "towering cerebellum" sign.[7] In addition to upward displacement, cerebellum also curves anteromedially around brainstem to give creeping cerebellum sign.

Although the characteristic features of Chiari II are of posterior fossa, multiple supratentorial findings are also mentioned in the literature.[3] Included in these are callosal dysgenesis, a beaked tectum, polymicrogyria, skull deformities(lemon sign), heterotopias, colpocephaly, prominent massa intermedia. The severity of PF is graded into mild, moderate and severe with mild being smaller than normal PF(<2mm) with a cerebella large enough to be identified and is not misshapen(figure 4).

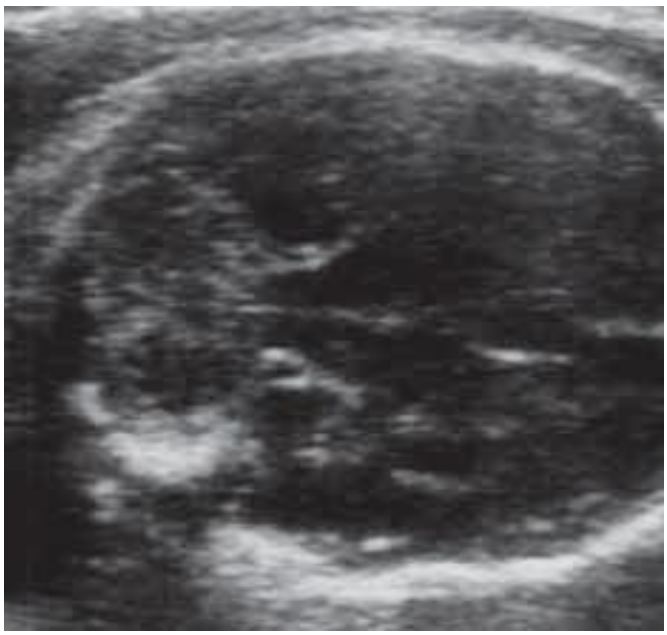

Fig 4: Mild deformity PF. Ultrasonogram shows small PF=1 mm with normal cerebellar hemispheres.

Moderate PF is present when PF is subjectively smaller with effaced cisterna magna and misshapen cerebellar hemispheres(banana sign), (figure 5)

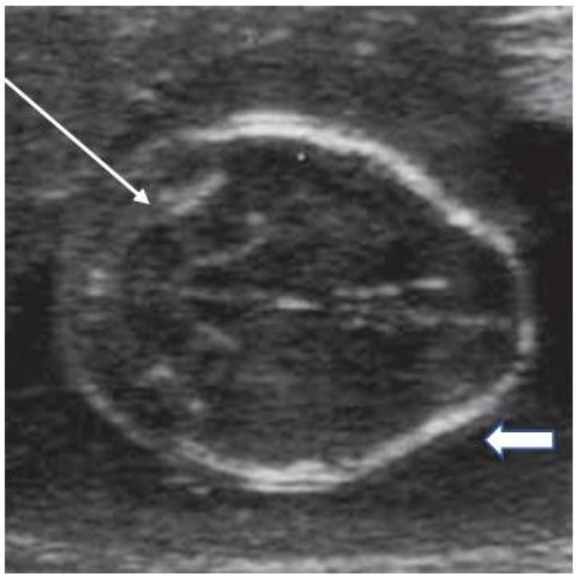

Fig 5: Moderate deformity PF. Ultrasonogram shows smaller PF with effaced cisterna magna with cerebellar hemispheres showing abnormal contour(Banana sign)(arrow). Note the presence of lemon sign(block arrow).

Severe PF deformity consists of very small PF with little or no cerebellar tissue visible (figure 6).

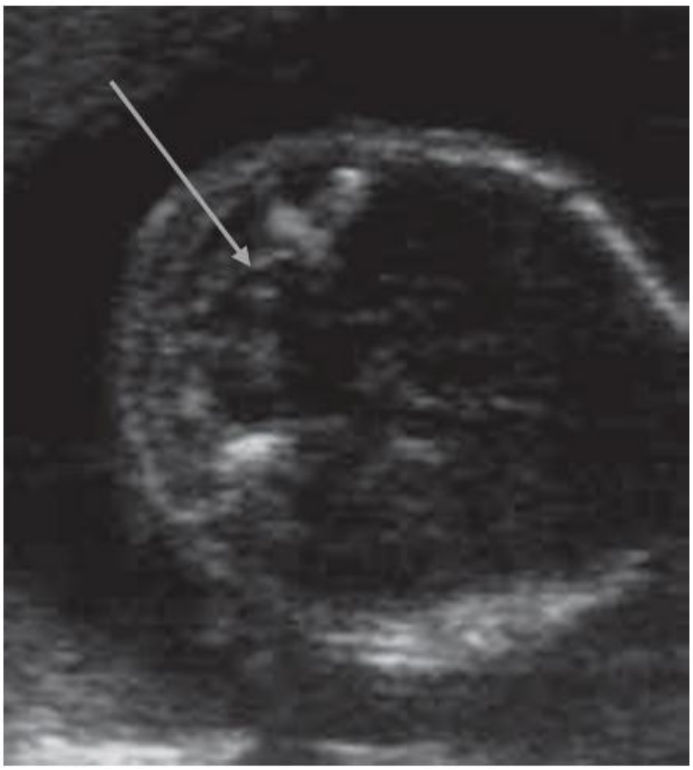

Fig 6: Severe deformity PF. Ultrasonogram shows very small PF with little identifiable cerebellar tissue(arrow).

For the differentiation from Chiari II, Chiari III malformation needs to have variable herniation of meninges or posterior fossa contents through low occipital or upper cervical bony defect(figure 7).

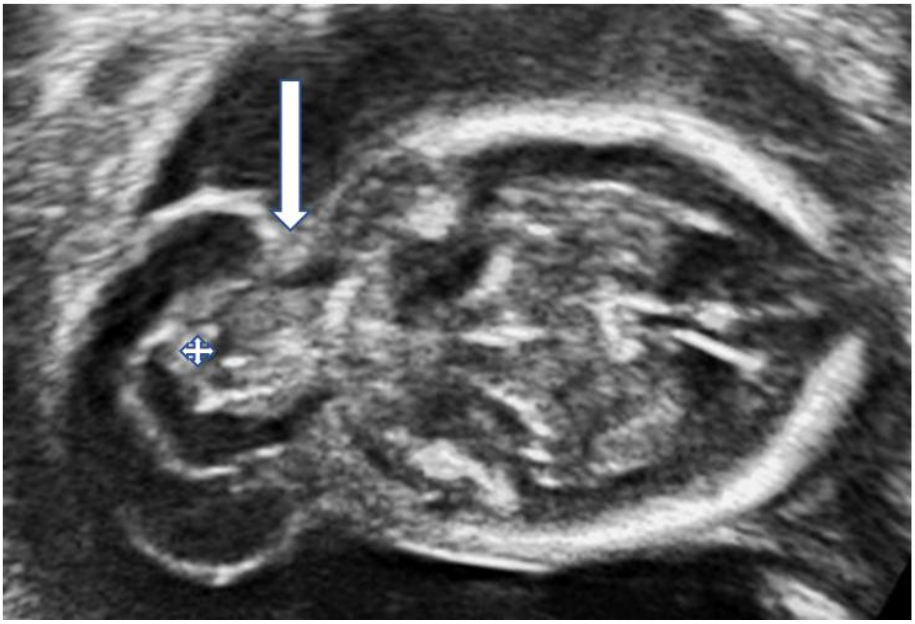

Fig 7: Bony Calvarial defect(arrow) with herniating posterior fossa contents.

CONCLUSION

The diagnosis of associated occipital or high cervical encephalocele is of paramount importance for multitude of reasons. It provides parents with an option to undergo karyotyping with possibility of termination of pregnancy. If the parents want to continue the pregnancy, adequate counselling and psychological preparation can be provided.

In our patient, timely diagnosis of Chiari III malformation with ultrasonogram provided ample amount of time for termination of pregnancy.

In conclusion, timely sonographic antenatal screening is emphasized as the primary method for detecting foetal congenital anomalies., thereby helping to make decision to offer further foetal karyotyping or termination of pregnancy.

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