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ARNOLD CHIARI TYPE II MALFORMATION: A CASE REPORT WITH PRENATAL SONOGRAPHIC FINDINGS

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ABSTRACT

Ultrasound imaging in the second trimester is essential for the identification of congenital anomalies. Chiari malformations, significant central nervous system (CNS) anomalies associated with high rates of mortality, can be diagnosed during pregnancy with either ultrasound or MRI. The diagnosis can include small posterior fossa, cerebellar herniation greater than 5 millimeters, hydrocephalus, various spinal defects, and the presence of myelomeningocele along with the lemon and banana signs. Early identification of these disabilities facilitates appropriate management of the pregnancy and care for the newborn. A 22-year-old woman, married for 10 months and a primigravida, underwent systemic and family history checks which revealed no significant findings. Her TIFFA scan at 22 weeks revealed several anomalies, including microcephaly, a defect at the back of the head with herniation of lower brain structures, hydrocephalus, and spina bifida with thoracolumbar myelomeningocele. This was diagnosed as Arnold Chiari malformation type 2 which is consistent with the findings. Her pregnancy was terminated at 24 weeks. Early diagnosis of such malformation helps to make decision to offer further fetal karyotyping or termination of pregnancy.

Keywords: Sonographic findings; Fetal anatomy; Cerebral hermination; Hyderocephalis.

INTRODUCTION

Second trimester ultrasonography is very valuable modality which gives valuable information about fetal anatomy and early detection of congenital anomalies. Among congenital CNS anomalies chiari malformations are most important with high mortality and morbidity . Early detection can help in termination of pregnancy or post partum follow up care. Diagnosis of Arnold chiari malformation is made with prenatal Ultrasound or MRI for evaluation of posterior cranial fossa. Small posterior cranial fossa with descending cerebellar herniation of more than 5mm from foramen magnum is considered diagnostic of chiari malformations. There are a variety of sonographic findings, some very specific for Chiari II malformations which include lemon sign, banana sign, hydrocephalus, posterior elements defect in spinal canal and associated myelomeningocele [1, 2].

CASE REPORT

A 22 year old pregnant women, primi gravida with 10 months of married life. No history of systemic illness, with no significant positive family history came for Targeted imaging for fetal anomalies (TIFFA) scan at 22 weeks of gestational age Sonographic findings showed multiple fetal anomalies including: small head measurements in mid trimester, small posterior fossa with downward displacement of the cerebellum, pons, medulla oblongata and fourth ventricle into the spinal canal through enlarged foramen magnum, obliteration of cisterna magna, mild obstructive hydrocephalus, spina bifida with thoracolumbar myelomeningocele.

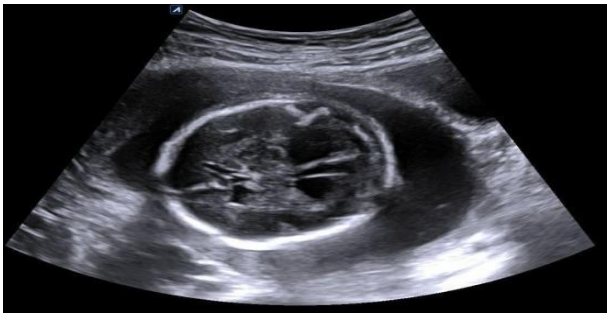

Fig 1: Sonographic image showing dilated lateral ventricles

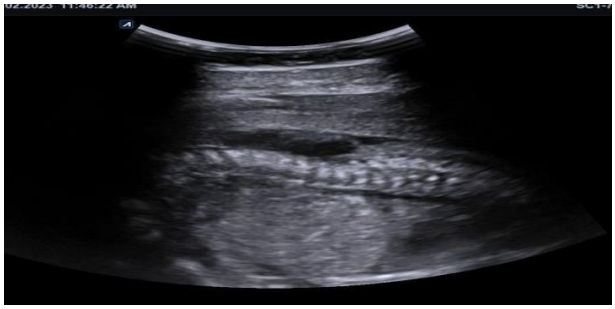

Fig 2: Sonographic image showing thoracolumbar myelomeningocele

According to these findings, Arnold chiari malformation type II was made and termination was performed at 24 weeks of gestation.

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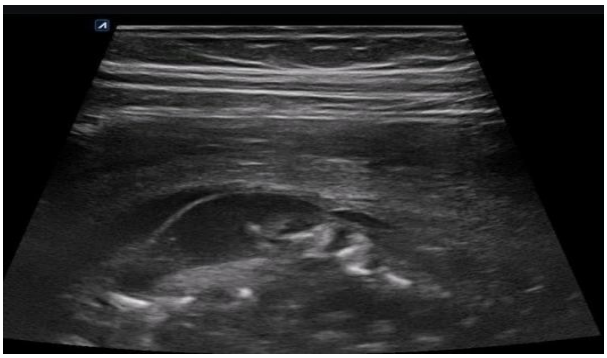

Fig 3: Sonographic image showing myelomeningocele

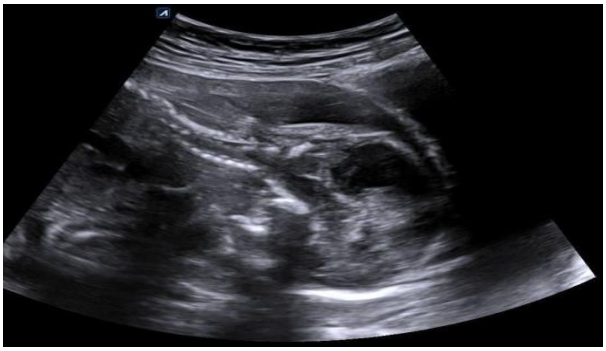

Fig 4: Sonographic image showing herniation of posterior fossa structures into cervical spinal canal through foramen magnum.

DISCUSSION

Malformations associated with dorsal induction like Arnold chiari malformations occur at 3rd -4 th gestational weeks [3]. There are four types of Arnold chiari malformations described in literature – Types 1 to 4. These can be confidently diagnosed by postnatal MRI evaluation. Type I: Herniation of caudal tip of cerebellar tonsil by >5mm, not associated with myelomeningocele. Type II: brainstem, 4 th ventricle and >5mm descent of caudal tip of cerebellar tonsil into spinal canal through foramen magnum with spina bifida. Features include hydrocephalus, Medullary kink, tentorial dysplasia and is almost always associated with neural tube defects like myelomeningocele. Type III: herniation of cerebellum associated with cephalocele or cranio cervical meningocele in which cerebellum and brain stem may be herniated. Type IV: cerebellar hypoplasia or aplasia with normal posterior fossa, with no hindbrain herniation [4]. Tubbs et al described two additional type of chiari malformation. Chiari type 0- Syringohydromyelia with distortion of contents in posterior fossa but without cerebellar tonsillar herniation. Chiari type 1.5- Caudal migration of brainstem and cerebellar tonsils often associated with syringomyelia. The feature of the Chiari II malformation which are most useful are the infratentorial findings, which include effacement of the cisterna magna, deformation of the cerebellum(banana sign), although other infratentorial abnormalities are commonly observed postnatally. Few studies have further divided the findings into mild, moderate and severe depending on the severity of narrowing of posterior fossa and alteration in morphology of cerebellum [5]. Descriptors in the literature ranging from" effacement of the fetal cisterna magna" to the banana-shaped cerebellum to the absent cerebellum have implied a continuum of severity of PF deformity. Few supratentorial abnormalities have also been described in literature which include abnormalities in tectal morphologic characteristics (fusion of the colliculi and upward deflection of the tectum result in prominent breaking and elongation of the tectum), altered shape of occipital horn that is pointed rather than rounded, corpus callosal dysgenesis, a small third ventricle, enlarged inter thalamic adhesions and colpocephaly. [6]. These are readily appreciated on post natal imaging by MRI. Rarely, visceral anomalies and other musculoskeletal abnormalities have been described The severity of posterior fossa (PF) deformity was graded to mild, moderate and severe. The PF deformity was considered mild when smaller than normal (<2mm) but identifiable; cisterna magna was present and the cerebella which was large enough to be easily identified, did not appear misshapen. A moderate deformity was diagnosed when the PF subjectively appeared somewhat small, the cisterna magna was effaced and misshapen cerebellar tissue could confidently be identified (banana shape). The PF deformity was considered severe when PF to be very small, the cisterna magna was affected and little or no identifiable cerebellar tissue was visible. The diagnosis of myelomeningocele in a fetus is important for many reasons. It provides the parents with an opportunity to consider pregnancy termination. Among parents electing to continue the pregnancy, adequate counseling and psychological preparation can be provided.

CONCLUSION

The ultrasonographic prenatal screening is emphasized as the primary method of assessment of the early fetal malformation. Early diagnosis of such malformation helps to make decision to offer further fetal karyotyping or termination of pregnancy.

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