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Chandana Ramesh
First Year Student,
Department of Anatomy, Shri
Atal Bihari Vajpayee Medical
College and Research Institute,
Bangalore, Karnataka, India

Dr. Meenakshi Parthasarathi
Professor and HOD,
Department of Anatomy, Shri
Atal Bihari Vajpayee Medical
College and Research Institute,
Bangalore, Karnataka, India

Corresponding Author:
Chandana Ramesh
First Year Student,
Department of Anatomy, Shri
Atal Bihari Vajpayee Medical
College and Research Institute,
Bangalore, Karnataka, India

A rare case of Arnold-Chiari syndrome in a primigravida and its outcome

Chandana Ramesh and Dr. Meenakshi Parthasarathi

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Abstract

The Arnold-chiari malformation is a spectrum of congenital malformations of central nervous system characterized by downward displacement of the parts of cerebellum, fourth ventricle, pons and medulla oblongata into the cervical spinal canal. It is one of the commonest causes for congenital hydrocephalus, it is associated with acromesomelic dwarfism for which exact etiology is not known. We are reporting a case of primigravida with full term gestation with type 2 Arnoldchiari malformation with CPD in labour. Emergency caesarean section was done. A live male 4.42 kg baby was extracted. Baby died on the 10th day.

Keywords: Arnold-chiari malformation, caesarean, ultrasound scanning, banana sign, lemon sign

Introduction

- Arnold-chiari malformation was first identified by Hans Chiari in 1891 in pediatric autopsy specimen. In legacy with name of his professor Dr. Arnold and his name Hans Chiari, the hind brain disorder is named as Arnold Chiari malformation.
- It has an incidence of 0.4 in 1000 live births and 1-2% of recurrence^[1]. Occurs more in female fetus than male. More prevalent in certain ethnicities (Britain, Wales, Ireland). Chiari classified CNS malformation into 4 types. Type 1 is usually asymptomatic, type 2 is the most common and seen in neonates and infants and associated with hydrocephalus and myelomeningocele^[2]. Type 3 is a high cervical encephalomenigocele^[3].
- Characteristic ultrasound features are frontal bone scalloping (lemon sign) and absent cerebellum or abnormal anterior curvature of the cerebellar hemispheres (banana sign)^[4].

Case Report

An unbooked 30year old primigravida with term gestation with Arnold Chiari Malformation Type II in fetus came with complaints of pain abdomen for which the pediatric opinion regarding the prognosis of the fetus was taken. On examination she was normotensive, pulse rate was normal, with no pallor or pedal oedema. Uterus was term size, acting with cephalic presentation and fetal heart rate was 140 beats per minute. On per vaginal examination cervix was well effaced, 5-6 cm dilated, membranes were absent, Vertex at -1 station with presence of CPD. So in view of CPD, emergency section was done and a live male baby of 4.42 kg was extracted with features of Arnold Chiary Malformation with myelomeningocele in the lumbar region. Intraoperatively there was maternal ascites and grade 3 meconium stained liquor. Baby got shifted to NICU and pediatrician counselled the patient attenders regarding the poor prognosis of the condition of the baby. Baby died on 10th day of life. The patient was discharged on 12th day.

Discussion

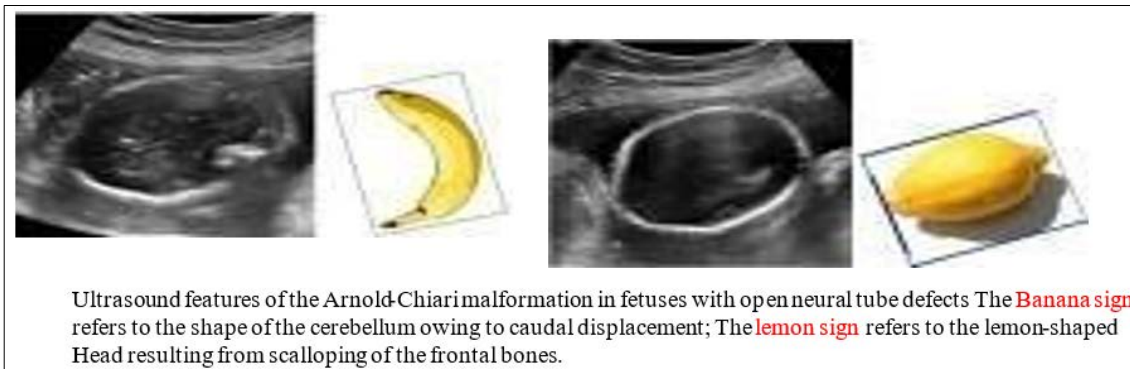
In legacy with name of his professor Dr. Arnold and his name Hans Chiari, the hind brain disorder is named as Arnold Chiari malformation. CNS abnormalities will be detected in routine second trimester anomaly scan. Chiari malformation is one among the CNS abnormalities. Normally the cerebellum and parts of the brain stem situated in an indented space at the lower rear of the skull, above the foramen magnum. When part of the cerebellum is located below the foramen magnum, it is called a Chiari malformation.

In Arnold Chiari Malformation, both cerebellar and brain stem tissue protrude into the foramen magnum and the cerebellar vermis may be only partially complete or absent.

- There are 4 types of CNS malformation
- Type 1 presents with only protrusion of cerebellar tonsils but intact fourth ventricle
- Type 2 presents with protrusion of cerebellum and brainstem into the spinal canal with spina bifida and associated neural tube defects
- Type 3 is rare type of brain herniation associated with cephalocele
- Type 4 is extreme cerebellar agenesis and downward displacement of cerebellum and brainstem^[5].
- 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” shape.
- 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” shape.

- Even though the exact etiology for this syndrome is not known, large meta analysis studies suggested that polymorphism of genes involved in folate metabolism result in neural tube defects^[6]. Danda S *et al.* study reported about a family siblings affected with acromesomelic dwarfism^[7]. haldar *et al.* described about the anesthesia concerns involved in patients with Acromesomelic dysplasia with associated hydrocephalus, Arnold Chiari malformation and syringomyelia^[8]. Urbizu *et al.* identified four genetic variants (located in the genes ALDH1A2, CDX1 and FLT1) to be associated with adult classic Chiari malformation type 1^[9]. As it is associated with severe morbidities, preconceptional folic acid supplementation is necessary. Its always advisable to terminate the pregnancy as and when it is diagnosed irrespective of the period of gestation.



Conclusion

Ultra sonography prenatal screening is a primary method of diagnosing fetal malformation. Early diagnosis helps to make decision on further continuation of pregnancy. preconceptional folic acid and vitamin B12 supplementation can prevent malformation.

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