

Case Report

IMAGING OF CHIARI TYPE I AND TYPE II MALFORMATIONS

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Abstract :

Chiari malformations (CM) are a group of posterior fossa abnormalities affecting the cerebellum, brainstem and the spinal cord. An in-depth understanding of the radiological findings is highly imperative for the early accurate diagnosis of CM in the developing fetus and the adult alike. We report here a case each of Type I and II CM presenting with the classical MRI imaging findings of CM Type I in a 38-year-old lady with complaints of a headache, seizures and loss of sensation in the lower limbs. And Arnold Chiari Type II during routine antenatal ultrasonographic screening in a 30-year-old primigravida at 17 weeks and 3 days of gestation respectively.

Keywords: Chiari malformations, posterior fossa abnormalities, MRI, ultrasonographic screening

INTRODUCTION

In 1891, Hans von Chiari described certain hindbrain abnormalities as postmortem findings in infants; these came to be known as Chiari malformations (CM). CMs are classified by the severity of the disorder and the parts of the brain that protrude into the spinal canal into types I, II, III, and IV.^[1,2]

Type I (Cerebellar tonsillar ectopia) is the most common form of CM. It involves the caudal herniation of the cerebellar tonsils into the foramen magnum, and is usually first noticed in adolescence or adulthood, often accidentally during a routine examination or in patients presenting with other complaints. Type I is the only type of CM that can be acquired.

Type II, (Arnold Chiari or classic CM) involves the extension of both cerebellar and brain stem tissue into the foramen magnum with a partially complete or absent cerebellar vermis. Type II is usually accompanied by a myelomeningocele, a form of

spina bifida, that causes the spinal cord and its protective membrane to protrude through a sac-like opening in the back. A myelomeningocele usually results in partial or complete paralysis of the area below the spinal opening. The term Arnold-Chiari malformation (named after two pioneering researchers) is specific to Type II malformations. Type II is the most common in neonates. This is usually associated with hydrocephalus and myelomeningocele.

Type III has the maximum fatality among the CM. The cerebellum and brain stem protrude or herniate, through the foramen magnum and into the spinal cord. The herniated cerebellar tissue can also enter an occipital encephalocele, a pouch-like structure that protrudes out of the back of the head or the neck and contains brain matter. The covering of the brain or spinal cord can also protrude through an abnormal opening in the back or skull. Type III causes severe neurological defects.

Type IV involves an incomplete or underdeveloped cerebellum—a condition known as cerebellar hypoplasia. In this rare form of CM, the cerebellar tonsils are located in a normal position but parts of the cerebellum are missing, and portions of the skull and spinal cord may be visible.

We report here a case each of Type I and II CM. Type I CM detected in an adult female presenting with complaints of a headache, seizures and loss of sensation of the lower limbs, while Type II CM in a primigravida on routine sonographic screening and lay forth the imaging findings that help in reliable diagnoses of the pathologies.

CASE REPORT

Case 1: A 38-year-old adult female presenting with complaints of a headache, seizures and loss of sensation in the lower limbs, on a diagnostic MRI scan, was found to have a caudal herniation of cerebellar tonsils (10 mm) into the cervical spinal canal below the level of the foramen magnum (ie. Mc Rae or basion-opisthion line) **Fig1(a,b)**. Crowding was noted at the level of the foramen magnum with a pointed configuration of the tonsils and effacement of the CSF space around the cervicomedullary junction **Fig1(c)** with an associated syringohydromyelia in the cervicothoracic region. Posterior osteophyte disc complex was also seen at the level of C5-C6 and C6-C7 indenting the thecal sac.

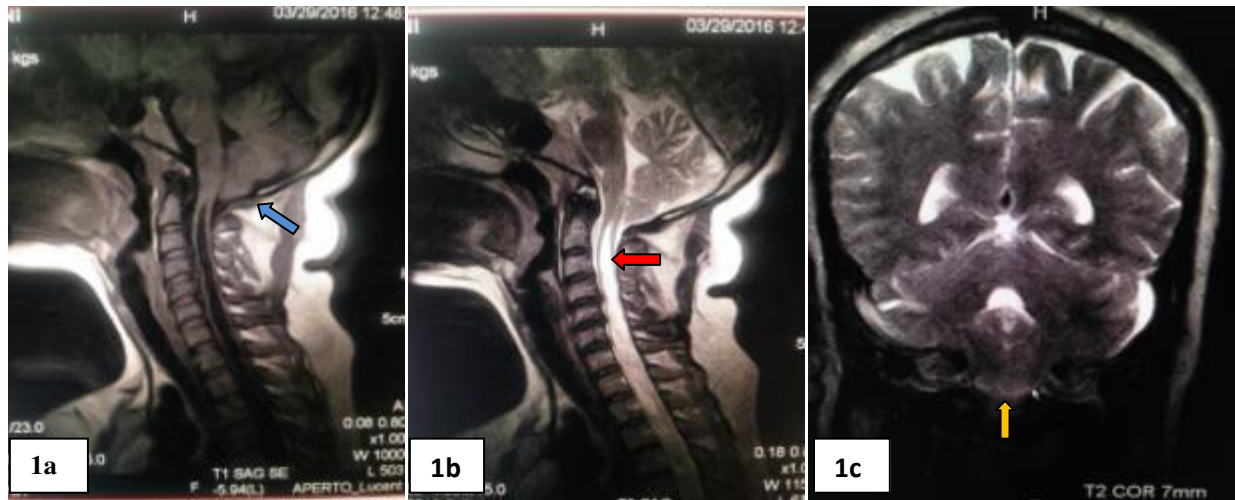


Figure 1 : T1 and T2W Sagittal images **1(a)** and **1(b)** show caudal herniation of cerebellar tonsils (10mm)(blue arrow) into the cervical spinal canal with associated syringohydromyelia in the cervicothoracic region(red arrow). T2 Coronal image **1(c)** shows crowding at the level of the foramen magnum with a pointed configuration of the tonsils (yellow arrow)

Case 2: A 30-year-old primigravida on routine sonographic screening at 17 weeks and 3 days of gestation, was found to have multiple fetal anomalies including bilateral inward frontal scalloping of skull called as ‘lemon sign’ **Fig 2(a)**, shallow posterior fossa with wrapping of cerebellum around the posterior brainstem known as ‘banana shaped cerebellum’ **Fig 2(b)** causing obliteration of cisterna magna and asymmetrical ventriculomegaly .**Fig 2(c)**

There is a cystic out-pouching with internal solid areas measuring 27 mm × 13 mm noted at the lumbar region suggestive of myelomeningocele **Fig 3(a)**. According to these sonographic findings, the diagnosis of Arnold-CM

(Type II) was confirmed and termination of pregnancy was advised. The sonographic finding myelomeningocele concurred with the post-mortem images **Fig 3(b)**. The diagnostic sonographic findings are inward frontal scalloping of the fetal skull ('lemon sign'), a shallow posterior fossa, an asymmetrical ventriculomegaly with dangling choroid, an abnormal anterior curvature of the cerebellar hemispheres ('banana sign') and lumbar myelomeningocele.

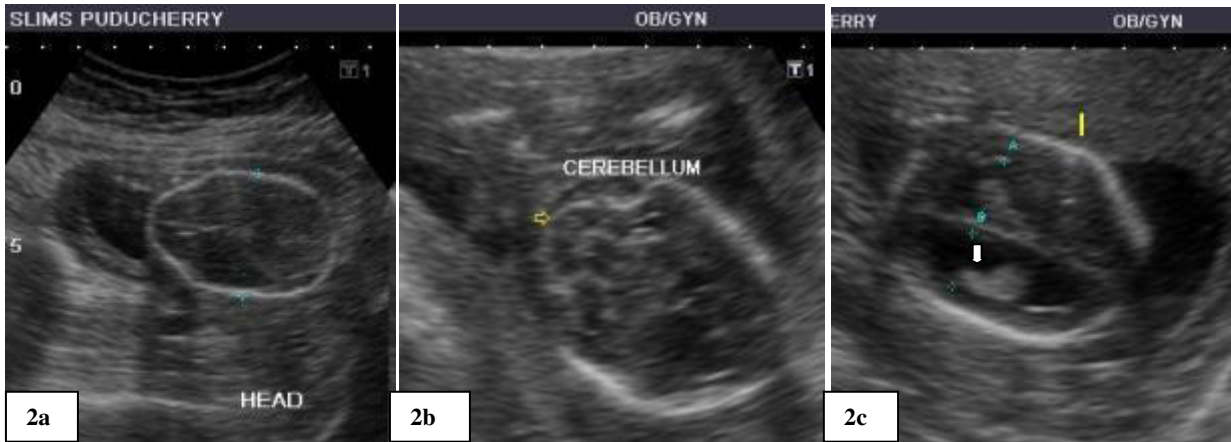


Figure 2 : Antenatal sonography of fetal skull and brain **2(a)** shows the 'lemon sign', **2(b)** displays the 'banana sign'(arrow) and **2(c)** shows asymmetrical ventriculomegaly with right dangling choroid (white arrow)

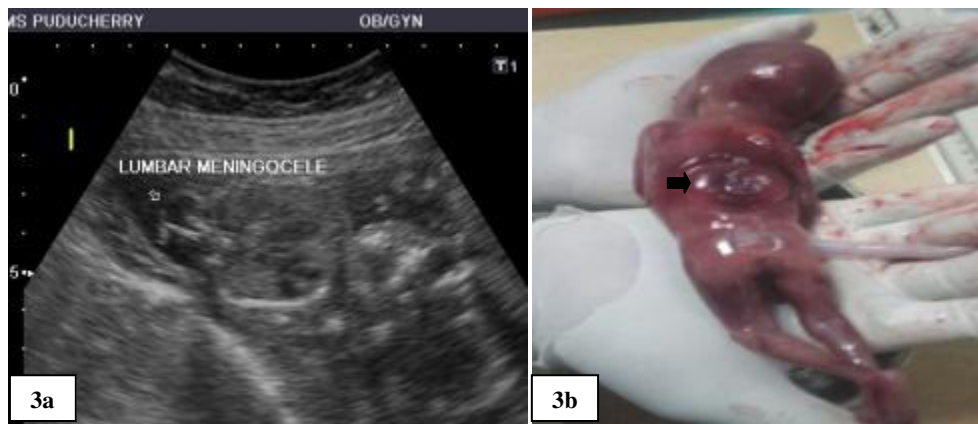


Figure 3: Antenatal sonography of fetal spine **3(a)** shows lumbar myelomeningocele (white arrow) and post-mortem image **3(b)** shows the myelomeningocele sac (black arrow) outpouching from the back in the same fetus.

DISCUSSION

Chiari Type I malformation is a congenital disorder recognized by caudal displacement of the cerebellar tonsils through the foramen magnum and into the cervical canal. Frequently associated findings include abnormalities of nearby bony and neural elements as well as syringomyelia. Cerebellar tonsillar ectopia is

generally considered pathological when greater than 5 mm below the foramen magnum. However, asymptomatic tonsillar ectopia is an increasingly recognized phenomenon, the significance of which is poorly understood.^[3]

Tonsillar ectopia, encompassing slight descent of the cerebellar tonsils and Chiari I malformations, are

disorders observed routinely in older children and adults and are believed to be an acquired form of the Chiari malformations. This entity is different from the other Chiari malformations in that hydrocephalus plays no role in its evolution. More likely it is a disorder of para-axial mesoderm, characterized by posterior fossa hypoplasia and content overcrowding, and not an embryologic anomaly of the neuroectoderm. The occipital exertional headache associated with this malformation can be observed in subjects who have new-onset tonsillar ectopia resulting from repeated lumbar puncture, idiopathic intracranial hypotension, lumbo-peritoneal shunting, or spontaneous development.^[4]

The pathophysiology of syringomyelia development is not fully understood. Current prevailing theories suggest that increased pulse pressure in the subarachnoid space forces cerebrospinal fluid (CSF) through the spinal cord into the syrinx. The here-proposed intramedullary pulse pressure theory instead suggests that syringomyelia is caused by increased pulse pressure in the spinal cord and that the syrinx consists of extracellular fluid. A new principle is introduced implying that the distending force in the production of syringomyelia is a relative increase in pulse pressure in the spinal cord compared to that in the nearby subarachnoid space. The formation of a syrinx then occurs by the accumulation of extracellular fluid in the distended cord.^[5]

Patients with objective brain stem or cerebellar signs had the largest mean tonsillar herniations. Patients with tonsillar herniations greater than 12 mm were invariably symptomatic, but approximately 30% of patients with tonsils herniating 5-10 mm below the foramen magnum were asymptomatic at MR imaging. "Incidental" Chiari I malformations are thus

much more common than previously recognized, and careful clinical assessment remains the cornerstone for proper diagnosis and management.^[6]

Our case, a 38-year-old female, presenting with complaints of a headache, seizures and loss of sensation of lower limbs, was found to have a cerebellar tonsillar ectopia of 10 mm below the McRae line (Basion-opisthion line), effacement of the CSF space around the cervicomedullary junction with associated syringohydromyelia in the cervicothoracic region.

The Chiari II malformation is a complex congenital malformation of the brain, nearly always associated with myelomeningocele, and the most common serious malformation of the posterior fossa include wrapping of cerebellum around posterior brainstem which is called 'banana sign', obliteration of the cisterna magna and other infratentorial abnormalities which are commonly observed postnatally. Supratentorial abnormalities includes hydrocephalus, corpus callosal dysgenesis, enlarged inter thalamic adhesions, a small third ventricle, a beaked tectum, polymicrogyria, heterotopias, non-visualization of the aqueduct, skull deformities (the 'lemon sign'), colpocephaly, stenogyria and ventriculomegaly. Chiari II malformation is best explained with the theory of McLone and Knepper, which allows the hindbrain disorder to be conceptualized as resulting from a normal-sized cerebellum developing in an abnormally small posterior fossa with a low tentorial attachment.^[7]

The prevalence of asymmetrical ventriculomegaly was more prevalent in Babcock study and often predominant in fetuses with moderate or severe posterior fossa abnormalities, especially later in the gestational period.^[8] Ventriculomegaly is considerably less common before than after 24 weeks

in fetuses affected with myelomeningocele. But in our case, asymmetrical ventriculomegaly, 'lemon sign' and shallow posterior fossa with 'banana sign' was seen at 17 weeks and 3 days of gestation.

The anomalies associated with posterior neural tube closure defects can no longer be considered secondary, but rather must be considered part of a spectrum of malformations caused by an unidentified primary insult to the central nervous system. The frequency and pattern of brain malformations associated with neural tube defects of some children with meningocele suggest that such malformations may seriously affect the intellectual outcome.^[9] The cranio-vertebral findings related to CM Type II are exclusively found in fetuses with myelomeningocele.

The diagnosis of myelomeningocele in a fetus is essential to consider termination of pregnancy.

Further evaluation of the spinal canal was performed in our case which showed lumbar myelomeningocele and was confirmed by the autopsy findings.

CONCLUSION

CM Type I encompasses a variety of disorders in relation to cerebellar tonsillar ectopia, presenting with an array of imaging findings on diagnostic MRI, which are specific in the diagnosis of the condition. CM Type II constitutes an interesting clinical entity with classical imaging findings which shows the need for ultrasonographic prenatal screening and helps to make a decision for early termination of pregnancy.

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