

## A rare case report of Arnold Chiari type III malformation

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### Introduction

Arnold-Chiari malformation (CM) is a congenital hypoplastic disease of the hindbrain which is characterized by spinal cord abnormality and hydrocephalus, due to some part of cerebellar prolapse(s) into the spinal cavity through the occipital defect [1]

Hans Chiari, Professor of Pathology at the German University in Prague, published a series on hindbrain herniations based on autopsy findings in 1891[2]. He described three classes of hindbrain anomalies, including Chiari malformation type III; he found this exclusively in patients with occipital and/or high cervical encephalocele, with herniated dysplastic posterior fossa contents, and other associated anomalies [3,4]. Arnold Chiari malformation type III (CM III) is the rarest of the Chiari malformations [5]

Newborn infants with CM III often suffer from respiratory failure, swallowing dysfunction, hypertonia, or amyotonia. Due to significantly respiratory failure associated with this type, the prognosis is known to be very poor [6].

### Case Report

A male baby was born at full term (37 weeks) through a caesarean section due to fetal distress weighing 2.1 kg, baby of non-consanguineous parents. Baby was kept in NICU for two days due to respiratory difficulty. There was neither history of any medicinal intake nor evidence of any maternal infection during pregnancy. No antenatal sonography was done. At birth a round swelling of size 3 cm \* 4 cm was present in occipital region, which was soft in consistency and covered by skin.

A MRI brain was done which showed evidence of Arnold Chiari type III malformation with low occipital and high cervical encephalocele. This encephalocele was projecting posteriorly and contained dysplastic occipital lobes and inferior cerebellum. Foci of haemorrhages were noted in the contents of encephalocele. Rest of the cerebellum appeared hypoplastic with small posterior fossa contents along with cerebellar tonsillar herniation as well. The brain stem appeared distorted and compressed with obliteration of posterior fossa CSF cisterns. Agenesis of corpus callosum was also noted with generalized effacement of cortical sulci and ventricles in supratentorial region as well suggestive of mild diffuse cerebral edema. In addition there was evidence of hypomyelination as well.

Visualized cervical and upper dorsal spine showed formation of syrinx in cervical and upper dorsal cord.

Patient relatives were explained about the prognosis and further management, but they refused for further management.

Patient again came to hospital after seven months with complaint of increase in size of swelling. There was history of delayed milestones. Patient was admitted, examined, investigated and managed surgically. On examination a swelling of size 15\* 10 \* 8 cm was present in occipital region which was soft, fluctuant, non transilluminant, thinning of overlying skin with shining surface. There were no other systemic disorders. Patient was investigated pathologically and radiologically. Pathology reports were within normal limits.

On Computed Tomography (CT scan) a bony defect was seen in the occipital bone measuring 34 mm (Transverse). A defect was also seen in the posterior arch of atlas vertebra. Herniation of cerebellum and both occipital lobes through the defect with large CSF density cystic component was noted suggestive of high cervical encephalocele. The encephalocele measures 13(CC)\*8.3(AP)\*8(TR) cm. Both lateral ventricles appeared mildly dilated. Fusion of body and posterior elements of C2 and C3 vertebra was seen. Overall features were suggestive of Arnold Chiari type III malformation.

On MRI a bony defect was seen in occipital bone and posterior arch of atlas through which herniation of cerebellum and both occipital lobes with large CSF intensity cystic component was noted which was suggestive of low occipital-high cervical encephalocele measuring 13.5(CC)\*9.2(AP)\*7.6(TR) cm. Cerebellum appeared hypoplastic with small posterior fossa contents. There was cerebellar tonsillar herniation noted. Distortion of brain stem was noted. Lateral ventricles were mildly dilated. There was thinned out corpus callosum noted.

Patient was managed surgically; Encephalocele repair with dysplastic brain removal was done. Specimen was sent for histology. Postoperative period was uneventful. Patient was discharged on post operative day 16. Histology report was suggestive of encephalocele.

### Discussion

Chiari malformations include a group of pathogenetically interrelated abnormalities amongst which Chiari I malformation is the mildest form characterised by caudal displacement of

cerebellar tonsils through foramen magnum into cervical canal. Chiari type II malformation is more complex characterised by myelomeningocele commonly in the lumbo-sacral region, hydrocephalus, hypoplasia of posterior fossa and associated supra tentorial findings. Chiari type IV malformation is characterised by hypoplasia or aplasia of cerebellum, small brain stem and abundant posterior fossa CSF [7-11].

Chiari type III malformation is associated with high cervical low occipital encephalocele. This malformation is the rarest of the Chiari malformations. The pathogenesis of this malformation is not fully understood. The theory of McLone and Knepper, which is the underlying patho-physiology of Chiari II malformation might be also, applied to Chiari type III malformation [12-13]. A lack of distension of embryonic ventricular system owing to the abnormal neurulation results in a hypoplastic posterior fossa and results in caudal displacement of cerebellum and brain stem. The low occipital osseous defect and associated cephalocele are probably developed due to failure of enchondral bone induction owing to the incomplete closure of neural tube or due to failure of complete fusion of ossification centres [14-15].

Chiari III malformation is characterised by herniation of posterior fossa contents which are cerebellum, sometimes brain stem and fourth ventricle through a low occipital or upper cervical osseous defect. Almost seventy percent cases show incomplete fusion of posterior arch of C1 and other cervical vertebrae.

The diagnosis of Chiari III malformation is based on neuro-imaging. CT is sensitive in the depiction of the bony defect but less sensitive in demonstration of herniated sac's contents. MR demonstrates the sac's contents with much greater accuracy which is necessary for the surgery. MR Angiography can help in screening of dural sinuses and major cerebral arteries relative to sac's contents. On USG diagnosis can be made as early as first trimester.

The treatment of Chiari III malformation is surgical closure of the cephalocele defect in order to prevent infection or rupture [7,8,12,15].

## Conclusion

We report an extremely rare case of Chiari III malformation associated with low occipital-high cervical encephalocele which

was managed with repair of malformation with dysplastic brain removal.

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