Case report

Arnold chiari type III andencephalocele, a rare and seriousassociation: a case report

ABSTRACT :

Arnold-Chiari type III ischaracterised by protrusion of the brain stem, cerebellum and lower part of the brain due to a cranial malformation. Encephaloceleis a rare congenital malformation in which the brain protrudes through a fissure in the skull. It occursduringpregnancywhen the neural tube of the foetusdoes not close completely. The association between Chiari malformation type III and encephalocele, althoughdescribed in the literature, remains rare, complicatingourunderstanding of theircommonpathophysiologicalmechanisms and their management.

We report the case of a femaleinfant ,admitted forseizurewithhypotonia and dysmophic syndrome. An MRI wasperformedshowingencephalocelewitharnoldchiari type III

The aim of this article is to understand and investigate the association between Arnold-Chiari type III malformation and encephalocele in order to develop more effective therapeuticstrategies.

Keywords: Chiari Malformation type III, MRI, Encephalocele ,Case report

INTRODUCTION :

The complex nature of Arnold-Chiari Type III and encephalocelerequirescareful recognition. It becomes important for all medicalfields to have a holisticview of theseneurologicaldisorders. Arnold-Chiari Type III, alsoknown as Chiari malformation, has a verylow occurrence in children. It is part of the spectrum of hindbrainherniationdisorders. The hindbrain contents, or the so-calledherniating parts, include parts of the mesencephalon, pons, medulla, cerebellum, and some portions of the fossaposterior's components and structures. At times, protrusion or herniation of hindbrain contents, meninges, and part of the skullisobservedthroughdefects in the cranium. The protrusions and the protruding parts are termedencephalocele. Such a neural tube defectmayresultfrommanyfactors, includinggenetic, neurological, environmental, and teratologicaletiology. Most cases remainasymptomatic for a longer duration beforepresentingwithtypicalhindbrainherniationsymptoms[1]

CasePresentation :

A patientborn 07/06/2023, admitted for seizure at the age of 1.5 years, with a history of pregnancyfollowed by vaginal delivery for a twinpregnancywith a birthweight of 2300 g, hospitalised at birth for hypotonia and dysmophic syndrome, an examinationwascarried out in the neonataldepartment: cardiac and abdominal ultrasoundwere normal. A transfontanellarultrasoundwasperformed, whichshowed an occipital encephalocelecompleted by a cerebral iris, suggesting an Arnold Chiari type III malformation with occipital encephalomeningocele and syringomyeliectomy.

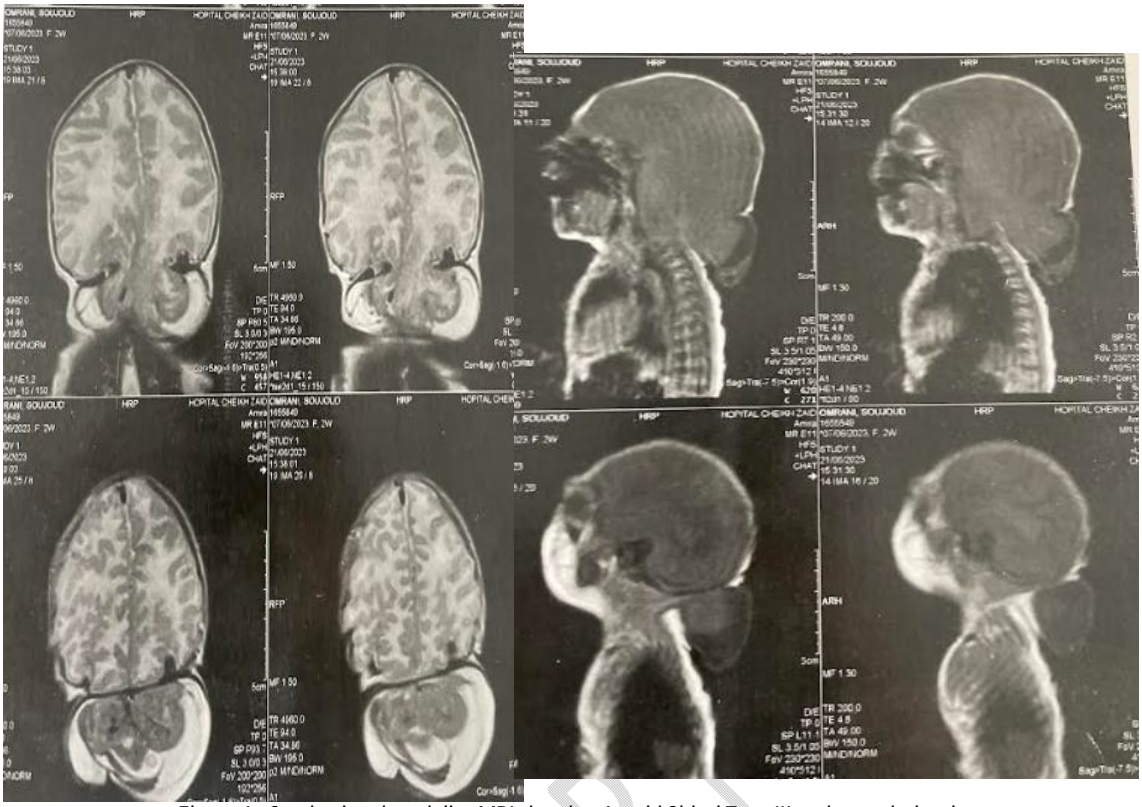

Figure 1 : Cerebral and medullar MRI showing Arnold Chiari Type III and encephalocele

operated on 11/2023, i.e. at the age of 5 months.

psychomotordelay: inability to hold the head up

Her medicalhistory dates back one week to the onset of a convulsive state consisting of the repetition of severalgeneralised tonic-clonic convulsions throughout the day, withoculardisgust, developing in the context of apyrexia, whichwas the reason for her consultation.

General examination on admission:hypotonic infant, apyrexia, absence of ocularpursuitwith facial dymorphia.

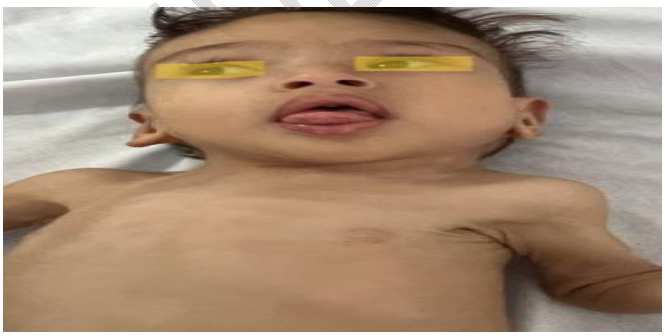

Figure 2 : Clinicalpresentation of a case of Arnold Chiari Type III

Weight and height and cranialcircumferencelessthantwo standard deviations.

Ophthalmologicalexamination:photomotor reflexes abolished, posture of visuallyimpaired.

EEG: diffuse slowing of background activitywith multiple wavepeaks.

Cerebr.al MRI: Arnold-Chiari type threemalformation.The patient wasdischargedwith sodium valproate and prescribedpsychomotorrehabilitation.

Discussion :

Encephalocele and AC Type III are malformations due to disrupteddevelopment. Encephaloceleisherniation of intracranial content through a bonydefect. It is more common in lessdeveloped countries (prevalence: 0.8 per 10,000 live births). Over 50% of encephaloceles are occipital and linked to embryologicvascular arachnoidbetween the 6th and 8th weeks of embryogenesis. 7-18% of encephalocele cases have severe mental and/or motorfunctiondisorders, depending on size, location, and brain tissue herniation. Mildintellectualprowessisseencompared to otherabnormalities. Previousstudies on headcircumference and encephalocele are limited.[2]

All healthcareworkers in the emergency, pediatric, and neurosurgicaldepartments, as well as the general population, must have accurate information about Arnold-Chiari type III and encephalocele for diagnosis and treatment. Clinicalassessment, medicalhistory, and neuroimaging are key tools for diagnosis. Ultrasoundisused for antenataldiagnosis, whileneonatal CT and pediatric MRI are used for postnatal diagnosis. Imaging findingscorrelatewithfunctionalimpairment and symptomaticonset. Differentialdiagnosisis important, consideringassociatedcranial anomalies. Accuratediagnosisis crucial for management, includingdeterminingintracranial volume and extent of bonedefect.[3]

Imaging iscritical for diagnosingtheseentities. MRI is the preferredmodality. Other options include CT and ultrasound. Imaging identifies important anatomicalfeatures, aidingtreatmentdecisions and prognosis. Radiologicalevaluationis key for accuratediagnosis. Modalitychoicedepends on presentationhistory. Severe cases require MRI, whilelesstypical cases can utilize MRI or ultrasound for supportive information.[4]

There are advantages and disadvantages to eachimagingmodality. CT and MRI are scarce and expensive for diagnosingencephalocele. Ultrasoundis the idealprotocol, with MRI alsobeingused. MRI is the mostcommon diagnostic method for encephaloceleassessment. Non-enhanced MRI or ultrasound can beusedinitially, followed by area-specificneuroimaging. The best modalitydepends on the clinical setting and suspicion level. Lateralencephaloceles show as hypodense areas on a CT scan. Babies shouldbe at safeweight and handledunderanesthesia for CT or MRI in the neonatalperiod.[5]

Severalsurgeriesrepair tissue layers to preventherniation and hydrocephalus. Techniques include dural repair, skull and dural repair, skull base reconstruction, fluidshunting, and preparation for definitiverepair. Initial treatmentstabilizes Arnold-Chiari Type III and prevents complications. Neurosurgery and symptom-based CSF diversion are used. Antibioticsreduce CNS infection risk. Postoperative monitoring assessesneurologicalstatus. CSF infection istreatedwithantibiotics and leaksmanagedwith drainage. Ultrasound and echocardiography check suture closure and cardiac complications. Long-term follow-up isrequiredwith multiple clinics.[6]

Low-grade Arnold-Chiari level 3 malformations and encephalocelesmay not requiresurgery. Therapyimprovesoutcomes. Referral to specialized centers for medication. Complementary alternative therapiesavailable. Integrated care at Neurotranslational Centers. Potential for science-based care.[7]

Conclusion :

the association of arnoldchiari type III malformation and encephaloceleremainsexceptional and serious. itsdiagnosisisrelativelystraightforward by clinicalexamination and imaging, especiallymri. managementismultidisciplinarywith a poor long-termprognosis.

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