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Case Study

Managing Chiari II Malformation in a Complex Pediatric Case: A Report

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ABSTRACT

Chiari II malformation (CM-II) is an innate CNS condition in which the medulla oblongata, pons, fourth ventricle, and cerebellum move lower inside the spinal canal. This abnormality is one of the leading causes of neonatal and infant death. CM-II diagnosis in the patient necessitates a thorough understanding of the findings. In this case report, we demonstrate Chiari malformation II, which is identified by an MRI of the brain and spine. This case also highlights the need for neuropsychological evaluation in CM-II in terms of providing guidance for psychoeducation and psychotherapy.

INTRODUCTION

The Chiari II malformation (CM-II) (Arnold-Chiari malformation) is a complicated congenital brain malformation that is almost invariably linked with myelomeningocele (see photos below). It is the posterior fossa's most frequent and significant abnormality.[1] This disorder manifests in the head, dural, cerebral cortex, and spinal column, with a horizontal migration inside the cortex, 4th ventricle, or cerebellum through the posterior spinal canal, together with pons and forth ventricle expansion, probably due to a small posterior fossa.[2,3] Differential diagnoses include spinal tumors, CM-I, and encephalocele. Other disorders to consider are 4th ventricular cell tumor, Lhermitte-Duclos sickness, rhombencephalon synapsis, and tectocerebellar dysgraphia-associated posterior encephalocele.[4]

Antenatal therapy for myelomeningocele lowers the requirement for ventricular shunting and improves motor results in newborns with CM-II. A study of 102 babies with open spinal dysraphism (OSD) indicated that in fetuses with severe CM-II who did not receive prenatal surgery, the majority (65.5%) had no improvement in the severity of cerebellum ectopia or Chiari grade postnatally. Most (81.3%) of fetuses that received in utero correction had postnatally resolved cerebellar ectopia.[5] CM-II has been shown to be completely reversible following postnatal myelomeningocele surgery. Beuriat *et al*. found that out of 47 individuals who were diagnosed with a CM-II verified before myelomeningocele surgery, only 28 (45.9%) remained CM-II following myelomeningocele closure. The reversible rate was 40.4%.[6]

CM-II is categorized by McLone and Knepper's hypothesis,

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which proposes that a cerebellum of typical size causes the hindbrain abnormality grows in an exceptionally small posterior region having a low tentorial connection.[7]

Clinical Report

A 3-month-old boy from Rajkot, Gujarat, diagnosed with Chiari II malformation (CM-II) was admitted to the hospital with symptoms including rupture of the swelling present on the back side, noisy breathing, problems eating or drinking, entering the respiratory system (aspiration), or rigidity in the arms.

Swelling has been present on the lumbosacral region since birth.

After tactile stimulation, the patient was kept on high-flow nasal cannula (HFNC) for 6 days and on O_2 with nasopharyngeal oxygen (NP) for 5 days.

Laboratory Investigations

MRI brain

Intraparenchymal hemorrhage with thinning of corpus callosum (dysgenesis), colpocephaly, and increased cerebellar tonsil herniation, so suggestion of CM-II.

MRI spine

Large defect $D_{11} - L_4$ with meningomyelocele, herniation of the spinal cord.

Other laboratory investigations are hemoglobin 8.3 g/dL (13–17 g/dL), WBC 20600 (4000–10000 cells/cm3), lymphocyte 67% (20–40%), HCT 25.5% (40–50%), serum creatine 0.34 mg/dL (0.7–1.40 mg/dL), and serum test 9.62 mg/dL (less than 0.6 mg/dL).

We concluded that birth asphyxia with meningomyelocele and hydrocephalus caused transient tachypnea in newborns (TTN) with hypoglycemia and premature rupture of membranes (PROM).

Neurosurgeons advise surgery at 6 months of age.

Pharmacotherapy

During hospitalization, the patient was treated with injections of Meropenem (190 mg, IV TDS), Vancomycin (70 mg, IV TDS), injection paracetamol (IV SOS), and multivitamin for nutrition. For a speedy recovery, patients were constantly under observation.

Discussion

A CM-II is an anatomical condition in which the foramen magnum, where the spinal canal connects the brain at the back of the skull, normally opens up to allow the brain to protrude out. The spinal cord and the brain are under strain as a result. Symptoms might range from minor to major. Most frequently, the issue is congenital, meaning it exists from birth.[8]

It is unclear exactly how CM-II develops. Although several explanations have been put forth, none fully account for all the characteristics. In general, it is believed that a malformation of the spine and brain structures during pregnancy is the main cause of CM-II. The 4th ventricle, medulla, and cerebellum are subsequently forced to enter the foramen magnum as a result. An in-utero CSF leak is assumed to be the cause of the concomitant myelomeningocele. CM-II is rather prevalent with a frequency of roughly 1 in a thousand live births. But figuring out the real frequency is challenging. In almost all instances, neural tube defects are frequently present, with myelomeningocele being the most frequent abnormality. Furthermore, the majority (about 95%) of myelomeningocele cases in newborns are related to CM-II. The kind of Chiari malformation that manifests most frequently in children is CM-II.[8]

The neuroanatomy shown on imaging is the main factor used to make the diagnosis of a patient with CM-II. The spinal fluid from the brain, cultured tissue, and blood do not yet contain biomarkers that can be utilized to validate the diagnosis. The most important modality for patient evaluation is magnetic resonance imaging (MRI) combined with neuroimaging. A CT scan can help determine the existence of CM-II in individuals who are unwilling to have an MRI. Fetal ultrasonography may occasionally be used to diagnose CM-II if fetal ventriculomegaly is present.[9] If spinal myelomeningocele is present, a detailed assessment is necessary to make the diagnosis of CM-II. The medulla and cerebellar vermis can be displaced downhill on an MRI, among other abnormalities that can help with the diagnosis of CM-II.[10]

The severity of the abnormality and the neurological problems it causes will determine how to treat CM-II. Surgical procedures are frequently needed. Several ways to treat hydrocephalus include sealing open neural tube anomalies and displacement of posterior fossa features. Some medical conditions, including apnea, respiratory failure, neurogenic bowel, and infant feeding issues, require nonsurgical treatment. Myelomeningocele frequently requires surgical intervention to be repaired. There is some evidence to support the benefits of doing the surgery on an unborn fetus.[11] For newborns who have a myelomeningocele diagnosis and no prenatal intervention, delivery should preferably be carried out in a medical facility with a level III neonatal intensive care unit. Prophylactic antibiotics are given, and the lesion is covered as part of the initial postpartum management. The lower back lesion should be operated on to close it during the first 72 hours.[12] The patient needs to be closely monitored after surgery in case hydrocephalus develops.[13]

It is necessary to periodically check for aberrant neurological function throughout life. A neurogenic bladder will almost always be present in myelomeningocele patients, which is significant. To lower the risk of kidney disease, intermittent catheterization is often necessary. A significant change in function may indicate an immediate neurological problem.[14]

Conclusion

Diagnosing the abnormality requires a detailed comprehension of this entity (clinical and patho-morphological signs) and magnetic resonance imaging (MRI). Children with CM-II have a significant death rate, although this early detection can help minimize the rate of brainstem compression symptoms and subsequent surgical decompression.

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