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Research Article

A CASE OF TYPE I ARNOLD CHIARI MALFORMATION WITH SYRINGOMYELIA

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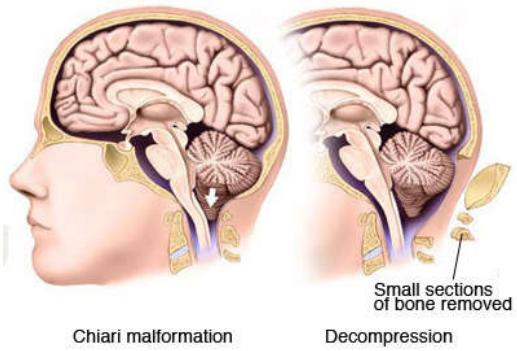
ABSTRACT

These Arnold Chiari Malformations Type 1 abbreviated as ACM are the structural defects of cerebellum and brain stem. It exhibits the downward displacement of the cerebellum at the base of the skull through the foramen magnum. Syringomyelia is the chronic degeneration and cyst formation on the spinal cord usually extended upto the Medulla Oblongata and Pons. Many of time it is associated with Ac malformations. [1] Women are three times more likely to have this congenital defect. Diagnosis of Arnold Chiari Malformation Type 1 is mainly based on clinical suspicion. Definite diagnosis requires assessment of patient's detailed history, neurological examination and medical imaging. [1,2] In this article we report a case of Arnold Chiari malformations Type 1 (ACM) with Syringomyelia in an 18 year old male patient.

INTRODUCTION

Arnold Chiari Malformations (Ac Malformation) often referred as Chiari malformations (CMs) are structural defects of cerebellum and brain stem in the base of the skull. It is exhibited by the downward extension of cerebellum tonsils through the foramen magnum (the opening at the base of the skull). It sometimes causes non-communicating hydrocephalus as a result of obstruction of cerebrospinal fluid (CSF) outflow. [1, 2]



Epidemiology

The prevalence of congenital Chiari I malformation was previously believed to be in the range of one per 1000 births, but is likely much higher.[3] Women are three times more likely than men to have a congenital Chiari malformation. The most common aetiology for ACM is the structural defect in the brain and spinal cord that occur during foetal development. Congenital Chiari malformations may develop when part of the skull is smaller than normal, which forces the cerebellum to be pushed down into the foramen magnum and spinal canal. This can be the result of genetic mutations or a maternal diet that lacked certain vitamins or nutrients such as vitamin B12 [4]. It can also be caused later in life if spinal fluid is drained excessively from the lumbar or thoracic areas of the spine either due to traumatic injury, disease, or infection. This is called acquired or secondary Chiari malformation. This causes pressure on the cerebellum and brain stem that may affect functions controlled by these areas and block the flow of cerebrospinal fluid (CSF)-the clear liquid that surrounds and cushions the brain and spinal cord. The most common type is Primary Chiari in comparison to secondary Chiari malformation.[1]

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Classification

Chiari malformations are classified by the severity of the disorder and the parts of the brain that protrude into the spinal canal.[1,5,6,7]

1. **Chiari malformation Type I:** It is the most common type. The cerebellar tonsils extend into the foramen magnum. It is usually asymptomatic diagnosed accidentally most of the time.
2. **Chiari malformation Type II:** In this case both the cerebellum and brain stem tissue are protruded into the foramen magnum. The vermis of the cerebellum may be missing or only partially formed. They suffer from more severe symptoms than Type I and it usually appear during childhood. It requires surgical management as it may cause life threatening complications.
3. **Chiari malformation Type III**: Type III is the rarest and most serious form of Chiari malformation wherein some part of the cerebellum and brain stem herniated through the skull along with surrounding membranes. In this case, some of the cerebellum and the brain stem herniate through an abnormal opening in the back of the skull along with surrounding membranes. The symptoms which appear during infancy can cause debilitating and life-threatening complications.
4. **Chiari malformation Type IV**: It exhibits an underdeveloped cerebellum (a condition known as cerebellar hypoplasia). The cerebellum is located in its normal position but parts of it are missing, and portions of the skull and spinal cord may be visible.

The common conditions associated with Chiari malformations are hydrocephalus, Spina bifida, Syringomyelia and spinal curvature.

Signs and Symptoms

Some people with Chiari Malformation may have no symptoms. Clinically the diagnosis is suspected by the presence of dizziness, muscle weakness, numbness, vision problems, headaches and problems with balance and coordination. Irritability when being fed, excessive drooling, difficulty in swallowing, a stiff neck, developmental delays are the common symptoms observed among infants. A detailed history collection of the patient, neurological examination and medical imaging will reveal the diagnosis of ACM.[1,3]

Magnetic resonance imaging (MRI) is considered diagnostic tool for Chiari malformation, since it visualizes the cerebellar tonsils and spinal cord as well as bone and other soft tissues. Prenatal ultra-sonography is also found helpful in detecting Ac Malformation. [1]

We report a case of Arnold Chiari malformations Type 1 (ACM) with Syringomyelia in an 18 year old male patient.

Case Report

An 18 year old male patient is presented with the chief complaints of headache since three years, walking difficulty since one year, ataxia and numbness in hands since one year. The patient also had a past history of perinatal asphyxia and delayed milestones. General and systemic examination revealed the following symptoms:

- Exaggerated DTR (Deep tendon reflexes).
- Pain in neck.
- Loss of sensitivity.
- Horizontal Nystagmus with Strabismus.
- Mild cerebellar atrophy.

Blood investigations were normal. Serological test for HIV was negative. The patient was afebrile with stable vitals and had fair GCS.

MRI Brain showed dilated 3rd and lateral Ventricles with associated mild thinning of corpus callosum. MRI Cervical Spine shows Arnold Chiari Type I Malformation (ACM) with syrinx formation.

Surgery is the predominant modality of management of Ac Malformation. The possibility of prognosis was discussed with patient and relatives and informed consent was obtained.

Surgical Management: Foramen Magnum Decompression (FMD) with C_1, C_2 Laminectomy with duroplasty was performed for the patient.

Postoperative Stage

Postoperatively the patient was given Injection Monocef (Ceftriaxone Sodium 500mg), Injection Amikacin (Amikacin Sulphate 100mg), Injection Pantop (Pantaprazole 40 mg), Inj PCM (Paracetamol 150 mg) and Injection Diclofenac (Diclofenac sodium 75mg).

Sr. No.	Trade Name	Generic Name	Action	Side Effects
1	Injection Monocef	Ceftriaxone Sodium 500mg	Antibiotic (Killing the bacteria by interrupting the bacterial cell wall formation.)	Vomiting Dizziness Heartburn Stomach pain Diarrhoea
2	Injection Amikacin	Amikacin sulphate 100mg	Aminoglycoside Antibiotic. It stops the growth of bacteria.	Bone pain Fever Headache Nausea Vomiting Diarrhoea
3	Injection Pantop	Pantaprazole 40 mg	Proton Pump Inhibitors (Antacid)	Headache and diarrhoea.
4	Inj PCM	Paracetamol 150 mg	Analgesic and antipyretic	Nausea, stomach pain and loss of appetite.
5	Injection Diclofenac	Diclofenac sodium 75mg	Analgesic and anti-inflammatory (NSAID)	Pain, dizziness, rise in blood pressure.

Outcome and Follow up

At the end of the 3-week treatment plan, the patient was asymptomatic with complete resolution of mechanical and neurological symptoms. The patient was re-evaluated by a Neurologist and Neurosurgeon, who acknowledged maximal medical improvement and complete resolution of the initial symptoms with no necessity for additional treatment, medication, or spinal surgery.

DISCUSSION

The treatments for Chiari malformation are surgery and symptomatic management. The presence of a syrinx gives specific signs and symptoms and these symptoms would definitely require decompressive surgery. They also develop neurological limitations.[6,7,8] In decompressive craniotomy, part of the lamina of initial cervical vertebrae and part of occipital bone is removed to relieve pressure. The flow of spinal fluid (CSF) may be improved by a shunt. A duroplasty may be performed as this surgery usually involves the opening of the dura mater and the expansion of the space beneath. The results seems to be excellent with improved brain stem function in majority of patients.[9]

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