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Case Report

Chiari Malformation- Report of 3 Cases

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Abstract

3 cases of chiari malformation type-I were presented with classical picture of spasticity, quadriparesis and dissociated sensory loss and were operated (Craniovertebral junction decompression by midline suboccipital craniectomy with removal of C1 arch and duroplasty). All the 3 patients were improved. Among them one patient recovered with some morbidity for postoperative other diseases.

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Introduction

Chiari malformations (CMs) are structural defects in the cerebellum, the part of the brain that controls balance. When the indented bony space at the lower rear of the skull is smaller than normal, the cerebellum and brainstem can be pushed downward.[1] These are not very common anomaly of craniovertebral junction. We have encountered six cases of Chiari malformation in two years. All of them were type-I. Three cases underwent operative treatment. Two patients recovered well. One patient recovered with some postoperative morbidity. This patient developed encephalitis one month after operation after initial recovery. She again developed enteric fever after recovery from encephalitis three months after operation. After all these ailments she got relief of morbidity with proper conservative treatment.

Case report

Case 1:

Mr. Akhter Mahmud of 38 years presented with neck pain, occipital headache, heaviness and weakness of all four limbs and burning sensation of upper thorax and both upper limbs. On examination it was found that he has spasticity of all four limbs, weakness of muscles of grade 4/5, all reflexes were exaggerated, plantar reflexes were extensor and bilateral Hoffman's test were positive. Dissociated sensory loss was found in cape like distribution. MRI of cervical spine showed type-I Chiari malformation along with Syringomyelia extending down to upper dorsal level.

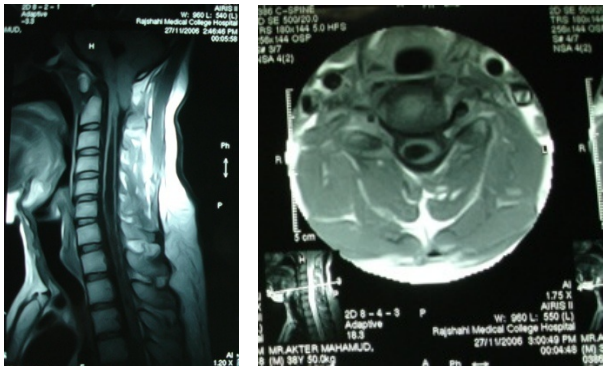

Fig: MRI showing Tonsillar herniation below the C1 arch and syringomyelia

Tonsil of Cerebellum reached down to C2 vertebra. Accordingly decision was made in favor

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of operation and midline sub-occipital craniectomy and removal of posterior arch of C1 done followed duroplasty with fascia lata from thigh. Postoperatively patient recovered uneventfully. Within one month all the symptoms improved except burning sensation which was relieved after three months.

Case 2:

Mrs. Mamtaz of 35 years presented with the complaints of occipital headache, neck pain, dizziness, fatigue and weakness all four limbs. On examination, there was spasticity and weakness (grade 4/5) of all four limbs, hyperreflexia and extensor plantar response and positive Hoffman's reflex bilaterally. Dissociated sensory loss was found in cape like distribution. MRI showed type-I Chiari malformation with syrinx extending down to mid dorsal region. Midline sub-occipital craniectomy and removal of posterior arch of C1 followed by duroplasty done. Immediately after operation patient recovered well, patient relieved of spasticity and weakness gradually recovering. But after 1 month patient developed severe headache, fever and vomiting and presented with depressed level of consciousness. For which she got admitted into the medical ward and was diagnosed as a case of encephalitis and was treated conservatively. 3 months after operation she again presented with high fever which was diagnosed as a case of enteric fever and was treated conservatively. After about 6 months of sufferings she recovered of her ailments.

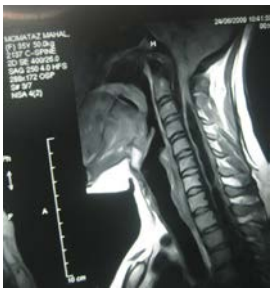


Fig: MRI showing tonsillar herniation below C1 with syrinx

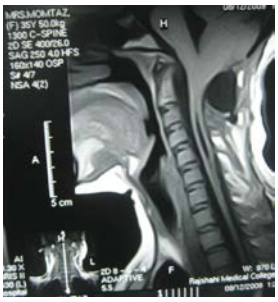
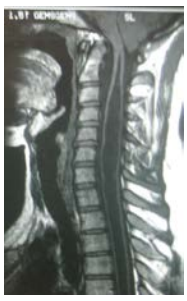


Fig: 6 wk. Postop MRI showing almost disappearance of syrinx

Case 3:

Mrs. Dolena 25 yrs. presented with severe spasticity and weakness of all four limbs. She also had occipital headache, vertigo, incomplete bladder emptying and dissociated sensory loss. MRI showed Chiari malformation type-I with severe syringomyelia. Tonsil reached down to C1 arch. Craniovertebral junction decompression done by midline suboccipital craniectomy, removal of C1 arch and duroplasty. Patient recovered well postoperatively. Her bladder control regained within 2 months. After 6 months her symptoms were relieved except mild sensory disturbance in the upper thoracic region mild spasticity.



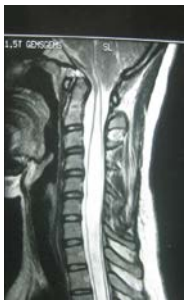


Fig: MRI of case 3, Syrinx reached down to mid dorsal region

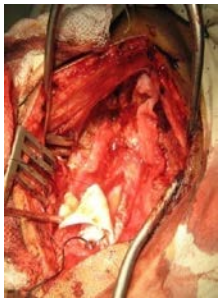


Fig: Duroplasty is going on

Discussion

Arnold–Chiari malformation, or often simply Chiari malformation, is a malformation of the brain. It consists of a downward displacement of the cerebellar tonsils through the foramen magnum (the opening at the base of the skull), sometimes causing non-communicating [2] hydrocephalus as a result of obstruction of cerebrospinal fluid (CSF) outflow.[3] The term "Chiari malformation" is preferred over the historical term "Arnold-Chiari malformation" due to the greater significance of the contribution of Chiari.[4] It can cause headaches, fatigue, muscle weakness in the head and face, difficulty swallowing, dizziness, nausea, impaired coordination, and, in severe cases, paralysis.[5]

An Austrian pathologist, Hans Chiari, first described these hindbrain malformations in the 1890s.[6] A colleague of Professor Chiari, Dr. Julius Arnold, later contributed to the definition of the condition.[7] The incidence is estimated to be in the range of one per 1000 to one per 5000 individuals.[8] The incidence of symptomatic Chiari is less but unknown.

The Chiari malformations consist of four types of hindbrain abnormalities, probably unrelated to each other. Majorities are type I & II [4] and Types III and IV are very rare.[9]

Type I Chiari malformation also known as primary cerebellar ectopia [10] a rare abnormality restricted to caudal displacement of cerebellum with tonsillar herniation below the foramen magnum and peg like elongation of tonsils.[4] Type II consists of caudally dislocated cervicomedullary junction, pons, 4th ventricle and medulla and usually associated with myelomeningocele.[4] Type III is the most severe form. Displacement of posterior fossa structures, with cerebellum herniated through foramen magnum into the cervical spinal canal often with high cervical or sub-occipital encephalomeningocele. It is usually incompatible with life. Type IV is cerebellar hypoplasia without cerebellar herniation.

Patients with CM1 may experience no symptoms. When symptoms are present, they usually do not appear until adolescence or early adulthood, but can occasionally be seen in young children. The majority of patients complain of severe headache and neck pain.[11] Three main patterns of clustering signs are found[12]- (a) Foramen magnum compression syndrome (22%)- Ataxia, corticospinal and sensory deficits, cerebellar signs and lower cranial nerve palsies (b) Central cord syndrome(65%)- dissociated sensory loss (loss of pain and temperature sensation with preserved touch and joint position sense and (c) Cerebellar syndrome(11%)- truncal and limb ataxia, nystagmus and dysarthria. Downbeat nystagmus is considereda characteristic of this condition.

Plain skull radiograph may show basilar impression, platybasia and cervical spine X-ray may show assimilation of C1, widened canal and agenesis of posterior arch of atlas. MRI is the diagnostic test of choice. It easily shows the classic abnormalities including tonsillar herniation and syringomyelia or hydromyelia [4].

Chiari I malformations that are asymptomatic should be left alone. There is no indication for "prophylactic" surgery on these. If the malformation is defined as symptomatic, or is causing a syrinx, treatment is usually recommended.[13]

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