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Original Research Paper

Neurosurgery

"A STUDY OF LONG TERM OUTCOME OF CHIARI MALFORMATIONS AFTER FORAMEN MAGNUM DECOMPRESSION: A CASE REPORT"

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ABSTRACT Chiari malformation (CM) is a pathology characterized by a downward displacement of one or both cerebellar tonsils through the foramen magnum. Clinical features are due to neurological compression and associated syrinx and these are suboccipital ataxia, cerebellar dysfunction and features of myelopathy. Patients having neurological worsening and syrinx need surgical management. We have described here a case of a young male, who presented with sensory disturbances and occipital headache. He was successfully treated by foramen magnum decompression and c1c2 laminectomy without opening dura and outcome is good. Patient is doing well after 13 years of surgery without any symptoms and recurrence.. We can conclude from this case that patients with chiary malformation with syringomylia after treatment with posterior fossa decompression with c1c2 laminectomy have good longterm outcome.

KEYWORDS : Chiari malformations, syringomylia, syrinx, syringohydromylia, foramen magnum decompression, pseudomeningocele, dissociative anesthesia

1. INTRODUCTION

Chiari malformation (CM) is a pathology characterized by a downward displacement of one or both cerebellar tonsils through the foramen magnum. The earliest description of this malformation was given by Dr. Hans Chiari(Austrian pathologist) which opened the door of detailed work and research on this topic.[1]

In Chiari malformation the most common presentation is suboccipital headaches and/or neck pain. Symptoms are exacerbated when asked to perform the Valsalva maneuver. Other common presentations include vision problems, dizziness, hearing loss, vertigo, gait ataxia, and generalized fatigue. Patients can present with isolated extremity pain or weakness.[2] Myelopathy classically presents with "dissociated sensory loss" (loss of pain and temperature sensation, preserved fine touch and proprioception) and motor weakness.[3] Cerebellar signs, including ataxia, dysmetria, and nystagmus, and lower cranial nerve deficits (IX, X, XI, XII CN) result either from direct compression of the cerebellum or medulla at the foramen magnum or from syringomyelia or syringobulbia.

There are two pathophysiological mechanisms for these clinical scenario: 1. Direct compression of neurological structures against the surrounding foramen magnum and spinal canal. 2. Syringomyelia or syringobulbia development. The obstruction of cerebrospinal fluid (CSF) outflow eventually results in syrinx development. Fluid-filled cavities (syrinx) develop within the spinal cord or brainstem, resulting in neurologic symptoms as the cavity expands.[10]

Chiari malformations are classified based on their morphology and severity of anatomic defects, typically noticed on imaging.[4]

Chiari I is the least severe, and often found incidentally. It is characterized by one or both cerebellar tonsils that project 5 mm below the foramen magnum, measured by a line drawn from the basion to the opisthion (McRae Line).

Chiari II consists of brainstem herniation and a towering cerebellum in addition to the herniated cerebellar tonsils and vermis due to an open distal spinal dysraphism.

Chiari III involves herniation of the hindbrain into a low occipital or high cervical meningoencephalocele.

Chiari IV is a rare variant that demonstrated severe cerebellar hypoplasia, similar to primary cerebellar agenesis.

There are other controversial reported classifications including Chiari 0, Chiari 1.5, and Chiari V. Chiari 0 is characterized by syringomyelia without hindbrain herniation, [5] while Chiari 1.5 is felt to be the progression of Chiari I with increased cerebellar tonsillar descent and some involvement of the brainstem.[6] Chiari V, the most severe variant, represents cerebellar agenesis with occipital lobe descent and herniation through the foramen magnum.[7]

Chiari I malformation is the most common type and occurs in approximately 0.5-3.5% of the general population with a slight female predominance (1.3:1). Chiari II occurs in 0.44/1000 births without gender predominance but can have a decreased incidence with folate replacement therapy by the mother in utero. [8][9]

Patients with Chiari malformation and who have no symptoms can be managed medically. Headaches and neck pain can be treated with muscle relaxants, NSAIDs, and temporary use of a cervical collar. However, studies show that while a headache and nausea may improve, in many symptomatic patients there will be no improvement in gait with medical management. Around 90% of patients with Chiari type I may remain asymptomatic even if they have syringomyelia.[11]

The main treatment for Chiari malformation is surgical with

the goal of re-establishing the CSF flow across the craniovertebral junction and relieving pressure on the cerebellum and hindbrain by decompressing the posterior fossa.[12]

Surgery is recommended for persistently symptomatic patients and confirmed tonsillar herniation. In the setting of asymptomatic tonsillar herniation, with or without syrinx, observation is recommended, unless symptoms develop.

Better surgical results are seen when surgery is performed within 2 years of symptoms onset. The standard surgical technique for Chiari I is a posterior fossa decompression. [13][14]This is obtained by a suboccipital craniectomy enlarging the foramen magnum, often in conjunction with C1, and possible C2, laminectomy. The dura may or may not be opened, with subsequent dissection of arachnoid adhesions if present. Depending on the available dural expansion and size of the posterior fossa, a duraplasty may need to be performed. The dural graft can be an autograft such as occipital fascia or tensor fascia lata (TFL) tendon, or artificial dura.[15] In the setting of a syrinx, a shunt can also be placed if decompression alone is not effective. Tonsillar cauterization may also be performed.

Initial surgical correction for Chiari II is the correction of the myelomeningocele, generally in the first 48 hours.[16] Closure of the spinal dysraphism can be done in a variety of ways, with either primary skin closure, myocutaneous flap, or fasciocutaneous flap, depending on the severity, involved layers, and the available adjacent tissue. The vast majority will eventually need a ventricular shunt for CSF diversion in the setting of hydrocephalus. If needed, a posterior decompression is performed later to allow suboccipital expansion.[17]

Chiari III follows a similar course to Chiari II. The occipital/high cervical encephalocele is corrected first, with resection of herniated contents, as these are typically non-viable, followed by dural closure and a cranioplasty.[18] A ventricular shunt is placed if the patient has concomitant hydrocephalus. The complications of Foramen magnum decompression include excessive bleeding, injury to vertebral artery intraoperatively, postoperatively cerebrospinal fluid leak, pseudomeningocele, infection, no relief of symptoms, craniocervical instability, paralysis, lower cranial nerve palsies and anesthesia complications.

In our institute, we have operated 55 cases of symptomatic chiary malformation with syringiomyia in last 6 years. There were 33 males and 22 females with male to female ratio 1.5:1. The mean age of the patients was 30 years and most common age group was 30-40 years. And we found very good outcome with very few complications on long term follow up. Here we have described a case of Chiary malformation with syringomylia treated with foramen magnum decompression with c1c2 laminectomy. We have followed the patient for 13 years for occurrence of any complication or recurrence. But there was no any complication or recurrence.

2. CASE REPORT

36 Years/male. came to opd for follow up examination. He was an operated case of post fossa decompression with c1c2 laminectomy on 3-10-2008. At that time patient had complaint of loss of sensations over left upper limbs, headache in occipital region since 1 year and weakness in left upper limb since 1 month.

On examination, patient was conscious, oriented, and vitally stable. On neurological examination he has poor hand grip on left side and power was 5/5 in all limb proximal muscles. There was sensory aesthesia on left side from c1 to D4 dermatome level for touch, temperature and pain sensations. While fine touch and vibration senstions were intact. The cranial neves, vision, extra ocular movements, tone, reflexes and gait was normal. There was no signs of cerebellar involvement or bowel bladder disturbances.

MRI brain was done and was suggestive of cerebellar tonsillar herniation and syringohydromylia extending from c1 to D6 level indicative of ACM type 1.

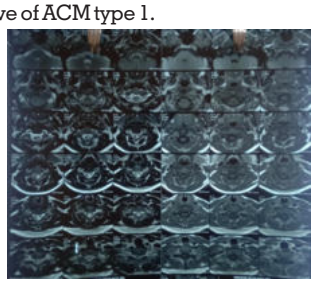

Figure 1. Preoperative MRI-1

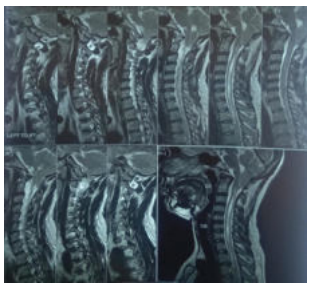

Figure 2. Preoperative MRI Image-2

After thorough pre operative necessary work up and anaesthetic fitness, post fossa decompression with c1c2 laminectomy done on 3-10-2008. Patient was operated under general anesthesia and in prone position with the help of bolsters and pillows with head kept over horse shoe. Midline suboccipital incision was taken extending from external occipital protuberance to c4 spine level. Posterior fossa craniectomy, foramen magnum decompression and c1c2 laminectomy was performed. Hemostasis was achieved. No evidence of csf leak was present. Closure was done in layers. Procedure was uneventful. Post-operatively, patient symptomatically improved with decreased headache, improved sensations and handgrip was improved on left side.

Patient was followed up regularly and patient had tremendous clinical improvement with only occasional tingling numbness and occipital headache. Follow up MRI showed decreased syringohydromylia and postoperative changes with no significant abnormality.

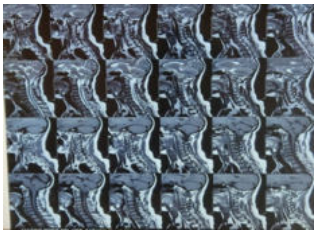

Figure 3. Follow up mri-1

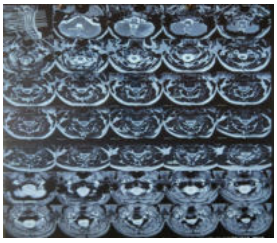

Figure 4. Follow up MRI-2

In follow up of 13 years, no any postoperative complication or recurrence was noticed. Patient has significant improvement clinically. With only minimal occasional occipital headache, tingling numbness. On clinical examination only slightly decreased pain sensations in affected dermatomes (c1-d4) with normal touch, temperature and pressure sensations.

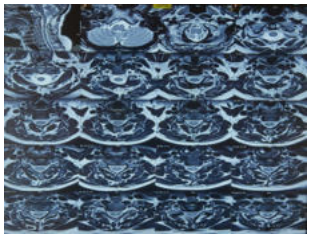

Figure 5. Follow up mri after 13 years of surgery-1

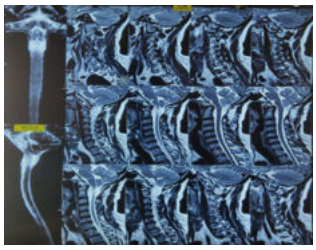

Figure 5. Follow up mri after 13 years of surgery-2

Figure 7. Postoperative Photograph on follow up visit showing clinical improvement

3. DISCUSSION

The Foramen magnum decompression has been historically considered to be the primary and gold standard operation for Chiary malformations. Herniation of cerebellar tissue due to presence of larger cerebellar mass in a relatively smaller posterior cranial fossa has been an important hypothesis of pathogenesis of Chiari malformation. Over the years, the extent of bone, dural and neural dissection has been reduced. Isu et al., recommended removal of only the outer dural wall.[19] Goel et al., suggested that only bone removal is sufficient for foramen magnum decompression and dural incision and duroplasty is not necessary.[20]

As per study of Giamatti et al [21], surgical treatment resulted in a long-term success rate of 84.2%. Two patients (5.2%) were unchanged after surgery, four patients (10.5%) showed recurrence of their symptoms. There was no mortality.

In the study of nasser ghandour et al[22], Patients with chiary malformation were divided into two groups: group I (32 cases) with syringomyelia and group II (14 cases) without syringomyelia. Posterior fossa decompression is recommended as the treatment of choice in adult Chiari I malformation with or without syringomyelia. They noticed that, the presence of syringomyelia predicts a less favorable response to surgical intervention.

But we have observed good clinical outcome on long term follow up in majority of patients with syringomylia as in case described above.

4. CONCLUSION

A case of Chiari malformation type I associated with syringomyelia was reported in a young male, who presented with sensory disturbances and occipital headache. He was successfully treated by foramen magnum decompression and c1c2 laminectomy without opening dura and outcome is good. Patient is doing well after 13 years of surgery without any symptoms and recurrence. It shows the good long-term outcome of foramen magnum decompression in chiary malformations with syringomylia.

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