Case Report

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Chiari Malformation-I with Syringomyelia: A Non-specific Presentation and Surgical Consideration

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Chiari Malformation-I is a congenital disorder characterized by the anatomical defect of the base of skull with tonsillar herniation (≥5 mm) below the foramen magnum, which is detected on MRI. It has a diverse range of symptoms with nonspecific presentation leading to the chances of misdiagnosis and untimely recognition of the disorder. Syringomyelia is the most common result of Chiari-I. The selection of surgical or non-surgical management depends upon the patient symptoms and the presence of absence of Syringomyelia. The objective of this case report is to give a broad perspective on Chiari Malformation-I from the symptoms and clinical findings obtained in a patient with Syringomyelia associated with Chiari Malformation-I and to discuss about the different surgical options as well as the psychological support required for the management of the condition.

Key Words: chiari malformation-I, posterior fossa decompression, syringomyelia, tonsillar herniation, syringo-subarachnoid shunt

Chiari disease (or malformation) is a congenital condition characterized by an anatomic defect of the base of the skull, in which there is narrowing of posterior fossa resulting in herniation of cerebellum and brain stem through the foramen magnum into the cervical spinal canal.Symptoms may vary between periods of exacerbation and remission. The diagnosis of Chiari Malformation–I in patients with or without symptoms is established with neuroimaging techniques.1,2 Chiari Malformation–I is classically defined as a cerebellar tonsillar herniation (≥5 mm) below the foramen magnum on sagittal Magnetic Resonance Imaging (MRI); it is estimated to be incidental, occurring in 0.6-1.0% of a given population.3 Over the past 3 decades, there have been a number of reports of Chiari malformation and Syringomyelia occurring either alone or combined,

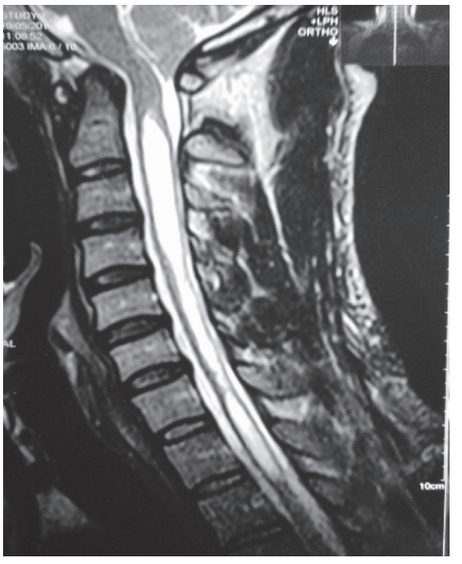

Figure 1: Sagittal view of MRI T2 weighted image showing hyperintense areas inside the spinal cord extending from C2 – D2

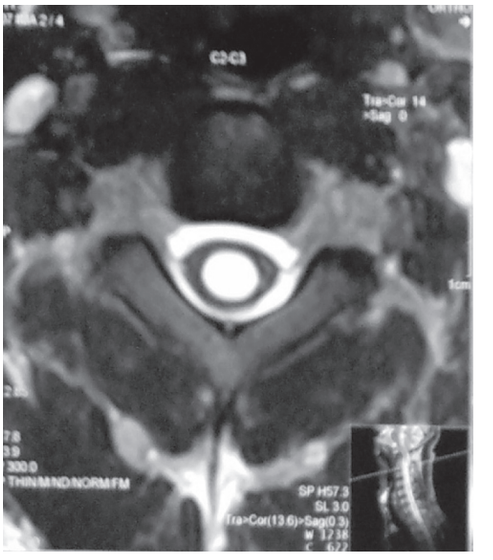

Figure 2: Axial view of MRI T2 weighted image showing hyperintense area inside the spinal cord

following cerebrospinal fluid (CSF) shunting from the lumbar subarachnoid space, birth trauma or tumors.3 Syringomyelia is a condition that results in fluid-containing cavities within the parenchyma of the spinal cord as a consequence of altered cerebrospinal fluid dynamics.4 The pathophysiology of Syringomyelia development is not fully understood. However, Current prevailing theories suggest that increased pulse pressure in the subarachnoid space forces cerebrospinal fluid (CSF) through the spinal cord into the syrinx. It is generally accepted that the syrinx consists of CSF5.The onset of Chiari syndrome symptoms usually occurs in the second or third decade (age 25 to 45 years), although it is commonly earlier in patients with Syringomyelia. Symptoms generally have an insidious onset and a progressive course. There is high clinical variability among patients, ranging from asymptomatic patients, patients with non-specific clinical manifestations, to patients with severe neurologic deficits1.We report a case of Chiari Malformation–I with Syringomyelia with a non-specific presentation presenting in a 9 months' period of time.

Case Report

A 34-year-old male presented with 9 months history of pain over left hand which was insidious onset and gradually progressive with tingling and burning sensation but he had no difficulty in performing physical activities. He also complained of numbness over left leg for 5 days. He had no history of trauma, sudden weakness, loss of consciousness, abnormal body movements, fever, paresis or paralysis of any part of the body. Bowel and bladder habit as stated by the patient was normal. He is non-hypertensive, non-diabetic and had no other chronic illness till date. He doesn't smoke but consumes alcohol occasionally. For the same problem, he had visited other hospital 20 days before presenting to us where he was advised for MRI C-Spine which showed Syrinx formation from C2-D2 with tonsillar herniation of 9 mm into the upper cervical spinal canal **(Figure 1, 2)**.

During presentation, his examination findings revealed decreased power (4/5) in all four limbs, however the grasp power was normal. There was no atrophy of thenar or hypothenar muscles. Pain and Touch sensation was decreased in the left side of his body with plantar reflexes bilaterally up going. All reflexes were exaggerated.

His CBC, ESR, Coagulation Profile, Immunology, RBS, ECG, RFT and LFT report showed no abnormalities.

He was then admitted and Posterior fossa decompression with laminectomy and Syringo-Subarachnoid Shunt under General Anesthesia was performed which released the blocked CSF pathway through foramen of Luschka & foramen of Magendie. Further management was done with IV fluids, Antibiotics, Analgesics, PPI, Multivitamins & other supportive measures. On the second and third post op day, patient had CSF leak from the surgery site so secondary suturing was done over the leak site. The problem persisted till the fourth day, so lumbar drain was kept, which prevented the CSF leak from wound site till fifth and sixth post op day. The leaking recurred till eighth post op day so revision surgery for posterior fossa repair was performed on ninth post op day.

Discussion

Chiari disease (or malformation) is a congenital condition characterized by an anatomic defect of the base of the skull, in which there is narrowing of posterior fossa resulting in herniation of cerebellum and brain stem through the foramen magnum into the cervical spinal canal. Based on the degree of herniation and the defect it manifests, it is divided into four types. However, Chiari type I malformation is undoubtedly the most frequent subtype. Chiari Malformation–I is classically defined as a cerebellar tonsillar herniation (≥5 mm) below the foramen magnum on sagittal Magnetic Resonance Imaging (MRI) **(Figure 3)**. Symptoms generally have an insidious onset and a progressive course. The onset of Chiari syndrome symptoms usually occurs in the second or third decade (age 25 to 45 years), although it is commonly earlier in patients with Syringomyelia. There is high clinical variability among patients, ranging from asymptomatic patients, patients with non-specific clinical manifestations, to patients with severe neurologic deficits.[1]

In a study done in 2009 among the patients diagnosed as Chiari Malformation-I, 37% were asymptomatic whereas 63%presented with some symptoms. The most common presenting symptoms were headache (55%), neck pain (12%), vertigo (8%), sensory changes (6%) and ataxia or poor coordination (6%). Other symptoms present at the time of diagnosis included leg weakness, tinnitus, hearing loss, dysarthria, loss of consciousness and scoliosis.6In very severe cases in which compression of the spinal cord or medulla oblongata occurs, symptoms of involvement of the motor or sensory pathways, or lower cranial nerves exist.[1]

The diagnosis of Chiari Malformation–I in patients with or without symptoms is established with neuroimaging techniques; the preferred technique is magnetic resonance imaging (MRI). MRI can also be used to study the volume of the posterior fossa and CSF flow dynamics. The diagnosis of Syringomyelia should be made by MRI of the complete spine (cervical, dorsal, and lumbar).[1]

In our case, there was presence of pain over his left hand with tingling and numbness indicating the radicular distribution which is usually absent in the patients with Syringomyelia. He had weakness of the limbs along with hyperreflexia along with loss of pain and touch sensation referring to the involvement of motor and sensory neurons by the syrinx. The presence of Chiari Malformation-I in MRI revealed the etiology of syrinx in the patient.

Regarding the modalities and treatment options, asymptomatic patients who are diagnosed of Chiari type I without Syringomyelia should not be considered as the candidates for surgery.In asymptomatic Chiari type I malformation with Syringomyelia, the opinion of

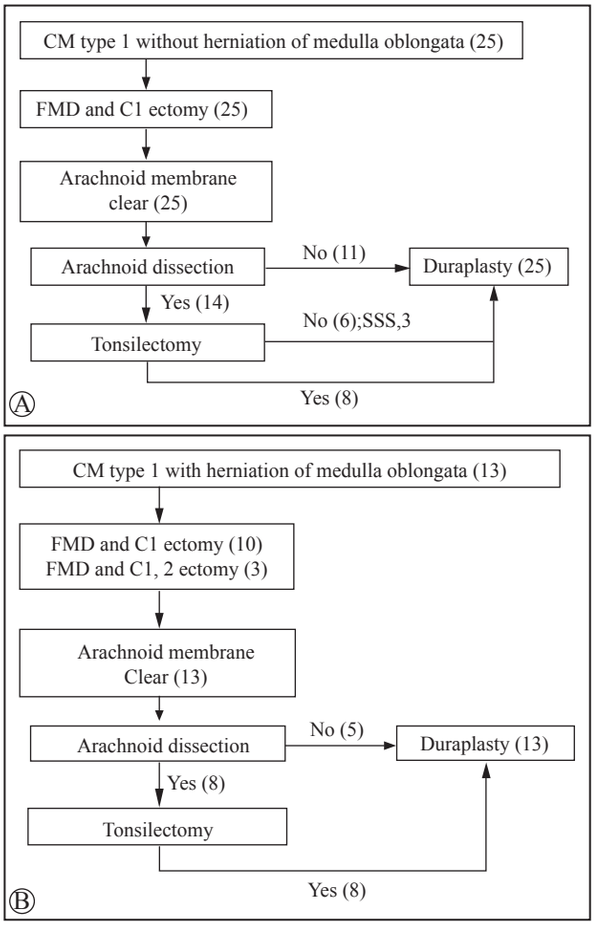

Figure 3: The algorithm of the surgical procedures. The surgical procedures for Chiari malformation (CM) type 1 without herniation of the medulla oblongata (A) and with herniation of the brainstem (B)7. FMD: Foramen Magnum DecompressionC1 ectomy: removal of the posterior C1 ringC2 ectomy: partial or total laminectomy of C2SSS: Syringo-subarachnoid shunt

neurosurgeons varies. In symptomatic patients, surgical treatment should be considered.[1]

In a retrospective study done in the cases of patients with Chiari Malformation-I who were surgically treated at an adult hospital from 1988-2013, Among 42 cases, Syringomyelia and clinical symptoms were improved in 32 (84%) and 31 (82%) patients, respectively. The sufficient restoration of the cisterna magna was the only significant factor. The reduction in Syringomyelia after restoration of the cisterna magna was not dependent on herniation of the medulla oblongata.[7] The algorithm of the surgical procedure along with the number of patients undergoing them are illustrated in Our patient was symptomatic with presence of Syringomyelia, so surgical option was chosen. The procedure we performed was Posterior fossa decompression with Syringo-subarachnoid shunt. The

patient symptoms were improved accordingly in the post-operative period.

As in all surgical procedures, decompression of the foramen magnum in Chiari malformation is not free from complications. Most of them involve CSF disorders, which are usually present in about 10% of patients. These include CSF fistula, meningitis, hydrocephalus, or the progression of Syringomyelia.

Postoperative relief of pre-operative pathologies was experienced in 83% of patients. Of the most common presenting symptoms, headache, neck pain and scoliosis, 12 and 17% respectively, were not alleviated postoperatively. However, the mortality rate, which is usually due to respiratory arrest in the immediate postoperative period or a serious sequela, should be less than 2%.1 Our patient also had CSF leak from the operated site as a complication of the procedure for which repair of posterior fossa had to be done. He recovered thereafter.

The debilitating disease like Chiari has a huge impact on the life of the patient as well the family members. The World Health Organization (WHO) defines health as "a state of complete physical, mental and social well-being, and not just the absence of disease or illness." The introduction of the social factor as an element that configures the state of well-being, together with the physical and mental factors, means that psychosocial care is needed to cope with a health problem like Chiari syndrome and Syringomyelia.

Conclusion

Chiari Malformation-I is a disease which is not so uncommon in the neurological world. However, the disease presents as thediverse symptoms and an irregular course, and these changing features are the factors which complicate most the timely diagnosis. A combined approach of symptoms, signs and a technical help from Magnetic Resonance Imaging (MRI) can help to reach a specific diagnosis of Syringomyelia.

The selection of treatment option following the diagnosis is the most critical decision of the neurosurgeon and neurologists. Though the procedure is not without complications, the balance between benefit and risk is to be evaluated before reaching a conclusion. In a patient who is symptomatic and MRI showing Syringomyelia, surgical option is always to be chosen.

It is necessary that professionals, especially primary care physicians, learn to identify the most characteristic symptoms of the most frequent craniocervical malformations (Chiari type I and Syringomyelia) in order to develop a diagnostic suspicion and refer appropriately patients for diagnostic confirmation.

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