

Incidental Detection of Arnold Chiari – I Malformation on MRI in a Case of Neck Pain

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ABSTRACT

Chiari malformations are spectrum of congenital hindbrain abnormalities. With advancing use of MR imaging, Chiari malformation is discovered with increasing frequency. A 19-year-old male presented with complain of the neck pain and occipital headache. No past history suggestive of trauma, fall or tuberculosis. Cranial nerve examination and higher function examination was within normal limits. Routine

blood investigations and chest X-ray were normal. Cervical spine showed partial occipitalization of the C1 vertebra. On MRI, partial occipitalization of C1 vertebra was seen with inferior cerebellar tonsillar herniation below the foramen magnum in upper cervical spinal with syrinx formation from C2 to L1 level. Patient was diagnosed as Arnold Chiari Type – I malformation based on the imaging features.

Keywords: Congenital hindbrain abnormalities, Occipital headache, Radiology

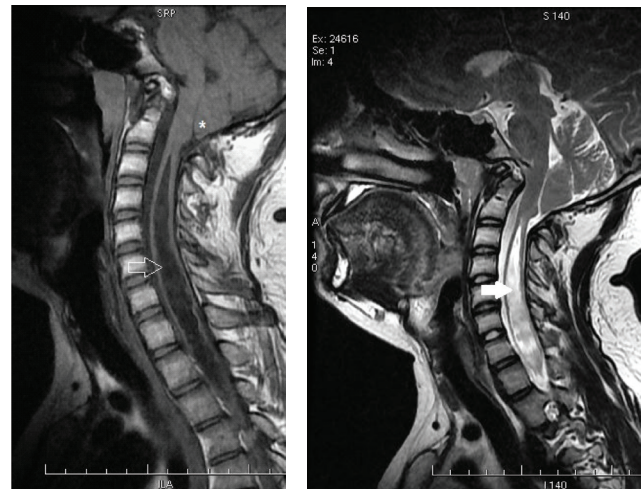
CASE REPORT

A 19-year-old male presented in the Medicine Department of the Shri Shayaji General Hospital, Vadodara with complain of the neck pain and occipital headache for the 15 days. Neck pain was not radiating or referred to the either upper limbs. No complain of fever. No past history of trauma, fall or tuberculosis. Family history was insignificant. Respiratory, cardiovascular and gastrointestinal examinations were normal. Cranial nerve examination and higher function examination was within normal limits. Grade-V power in bilateral upper and lower limbs. Tone and reflexes were normal. Oral cavity examination revealed normal. No features suggestive of tonsillitis and pharyngitis.

Haemoglobin (Hb) was 12.5 gm%, Total white blood cell (WBC) counts were 6700 cells/mm³, ESR and rest of the routine blood investigation were within normal limits. Chest X-ray did not reveal any abnormality. Partial occipitalization of the C1 vertebra with preserved cervical lordosis was present in cervical spine X-ray. Then physician advised to do Magnetic Resonance Imaging (MRI) cervical spine.

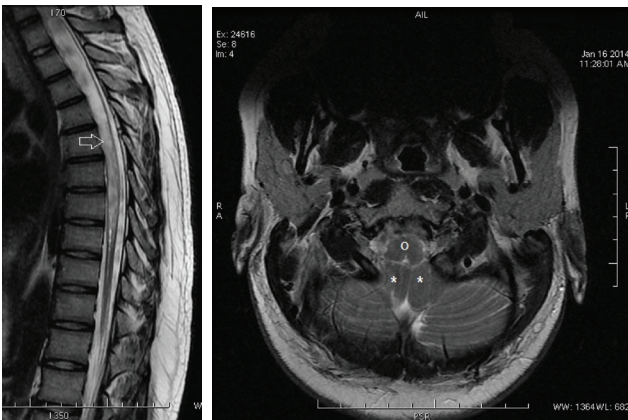
On MRI cervical spine, partial occipitalization of C1 vertebra with inferior cerebellar tonsillar herniation measuring 9 mm below the foramen magnum in upper cervical spinal. Focal longitudinal well defined, extensive abnormal signal, hyperintense on T2W and hypointense on T1W images with prominent CSF flow void within, was seen in central part of spinal cord extending from C2 to L1 level causing expansion

of the spinal cord and peripheral thinning of the parenchyma suggest syrinx formation. However, anterior atlanto-dens



[Table/Fig-1]: Sagittal T1W image of cervical spine showing inferior cerebellar tonsillar herniation (asterisk) in upper cervical spinal, measuring 9.0 mm below the level of foramen magnum. Focal longitudinal well defined, extensive hypointensity on T1W images seen in central part of spinal cord extending from C2 to L1 level (open white arrow) causing expansion of the spinal cord and peripheral thinning of the parenchyma

[Table/Fig-2]: Sagittal T2W image of cervical spine showing inferior cerebellar tonsillar herniation in upper cervical spinal below the level of foramen magnum. Focal longitudinal well defined, extensive hyperintensity (solid white arrow) corresponding to hypointensity on T1W images with prominent CSF flow void within, causing expansion of the spinal cord and peripheral thinning of the parenchyma suggestive of syrinx formation



[Table/Fig-3]: Sagittal T2W image of thoracic spine showing syrinx formation extending from C2 to L1 level with prominent CSF flow void within (open white arrow).

[Table/Fig-4]: Axial T2W image at the level of foramen magnum. Note crowding of foramen magnum by the ectopic cerebellar tonsils (asterix) and the medulla (open white zero). Also note the absence of cerebrospinal fluid.

interval and cervical lordosis were preserved. No significant narrowing of spinal canal or compression of spinal cord at cranio-vertebral junction [Table/Fig- 1-4].

Patient was diagnosed as Arnold Chiari Type – I malformation based on the imaging features. Then patient was referred to the higher center for the further management.

DISCUSSION

Chiari malformations are spectrum of congenital hindbrain abnormalities. It usually involves the structural relationships between the cerebellum, brainstem, the upper cervical cord and cranial bones. There are four types of Chiari malformations I, II, III and IV. Chiari malformation is listed as a rare disease but with advancing use of MR imaging, Chiari malformation is discovered with increasing frequency. Prevalence rates of Chiari I is 0.1-0.5% with a slight female predominance [1]. Chiari II is associated with myelomeningocele. Studies suggest linkage to chromosomes 9 and 15 for the Chiari I [2].

Chiari type I is that it originates as a disorder of para-axial mesoderm, which leads to the small posterior fossa. With the development of the cerebellum within this small posterior cranial fossa results with herniation of the cerebellar tonsils [3].

In 23 - 45% of cases there are also skeletal anomalies like platybasia, basilar invagination, atlanto-occipital assimilation, Sprengel deformity, Crouzon syndrome, Hajdu-Cheney syndrome and Klippel-Feil syndrome [4]. Symptoms develop as a result of compression of medulla, upper spinal cord, cerebellum and disruption of CSF flow through foramen magnum. Myelopathy and lower cranial nerve dysfunction

occur due to compression of cord and medulla. Ataxia, dysmetria, nystagmus, and dysequilibrium occur due to compression of cerebellum. Disruption of CSF flow through foramen magnum leads to the most common symptom, pain. Coughing and Valsalva manoeuvre aggravates headache and neck pain in Chiari I. Hydrocephalus occurs less frequently.

Normally the foramen magnum is an oval-shaped opening in the occipital bone, surrounded anteriorly by the clivus, laterally by the occipital condyle, and posteriorly by the squamous portion of the occipital bone. Through which the medulla traverses and merges with the cervical cord. Cisterna magna normally forms a large CSF cushion behind the medulla within the foramen magnum. This CSF cushion is replaced by cerebellar tonsils in Chiari I malformation.

MRI is the non invasive useful imaging modality for the diagnosing Chiari malformation. It shows the anatomy of the cranio-cervical junction and provides useful information about associated abnormalities, like syringomyelia and hydrocephalus. Symptomatic Chiari I is differentiated from cerebellar ectopia by the CSF flow analysis through foramen magnum [5]. It is also helpful to predict surgical response. Lab studies are not much valuable for diagnosing Chiari malformations.

Tonsillar ectopia is an asymptomatic condition. It found incidentally where the tonsils protrude through foramen magnum by less than 3-5 mm.

Conservative treatment option is for the patients with minimal symptoms without syringomyelia. Mild neck pain and headaches can be treated with analgesics, muscle relaxants, and use of a soft cervical collar. Symptomatic patients, such as those with lower cranial nerve dysfunction, myelopathy, syringomyelia, cerebellar symptoms, or severe post-tussive suboccipital headaches required surgery [6]. However, syrinx persists after surgery at an average rate of 6.7% [6]. Main goals of surgical treatment are decompression of Cervico-medullary junction and restoration of normal CSF flow.

Prognosis after surgery is good and depends on the extent of preoperative neurological deficits. Patients without the neurological deficit have excellent outcome.

CONCLUSION

Chiari malformations are spectrum of congenital hindbrain abnormalities. Chiari malformation is discovered with increasing frequency by the use of MR imaging. Some skeletal anomalies like platybasia, basilar invagination and atlanto-occipital assimilation may be associated with it. Conventional radiography has no role in the diagnosis. MRI is the investigation of the choice for diagnosing Chiari malformation. Conservative or surgical treatment decided based on the symptoms and complications. Prognosis after

surgery is good. Excellent outcome is seen in the patient without neurological deficit.

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