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Chiari I malformation with holocord syrinx

A 23-year-old man presented with a 1-month history of headache and numbness of the left arm and hand. Neurological examination showed hypesthesia in the left arm. Magnetic resonance imaging of the spine showed peg-shaped herniation of tonsils 9 mm below the foramen magnum (fig 1A), with altered signal intensity in the medulla (fig 1B). There was no hydrocephalus. T1W and T2W screening of the whole spine showed holocord syringohydromyelia (fig 2A,B). No focal intraspinal mass was seen. Chiari I malformation with a holocord syrinx was diagnosed.

Chiari I malformation is traditionally defined as inferior displacement of the cerebellar tonsils and, sometimes, the inferior vermis through the foramen magnum into the rostral cervical canal. Between the ages of 5 and 15 years, descent of upto 6 mm is not considered pathological. However, in older patients, tonsillar protrusion ≥ 5 mm is associated with a marked rise in the onset of clinical symptoms.¹

The aetiopathogenesis has been a focus of considerable controversy. It may be congenital or acquired. The causative factors include underdevelopment of the posterior fossa, malformation of the craniocervical junction, and changes in intracranial pressure.

The clinical presentation is varied. The patients may present with headache or oculomotor disturbances (blurred vision, oscillopsia). Otoneurological complaints (dizziness, tinnitus), cranial neuropathies (dysphagia, dysarthria) or cerebellar abnormalities (dysmetria, tremor) may be the other modes of presentation. Potentially serious autonomic disturbances are also common: sleep apnoea, respiratory failure, syncope and even sudden death. Skeletal anomalies that may be seen in association with Chiari I malformation include basilar impression, atlanto-occipital fusion, atlanto-axial assimilation, Klippel Feil deformity and scoliosis.^{2,3}

The concurrence of Chiari I malformation and syringohydromyelia has been estimated at between 25% and 65%. This results from obstruction of cerebrospinal fluid circulation in the cisterna magna. The presence of a syrinx is more commonly associated with symptoms.³

The various surgical treatment options can be broadly categorised into four groups: (1) decompression of hindbrain malformation by suboccipital craniectomy and upper cervical laminectomy; (2) laminectomy and syringostomy; (3) terminal ventriculostomy; and (4) percutaneous aspiration of the syrinx. The first group is the most common surgical option used in cases of Chiari I malformation and syringohydromyelia.²

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Figure 1 (A,B) Consecutive sagittal T1W images show herniation of peg-shaped cerebellar tonsils 9 mm below the level of the foramen magnum. The visualised cervical cord shows mild expansion with hydrosyringomyelia. The medulla shows altered signal intensity, distinct from the cord syrinx, probably due to pressure necrosis.

Figure 2 (A,B) Sagittal T1-weighted image of the whole spine shows the syrinx involving the entire cord.

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