Cranio-cervical junction malformation: Our series of Chiari Type I

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Background: The Chiari malformation (CM) is a rare congenital nervous system malformation that consists of the herniation of the cerebellar tonsils below the foramen magnum that may causes severe disability due to its neurologic symptoms. The most common type of CM is type 1. It has a prevalence in the general population between 0.24% and 3.6%. However, only 32% of the incidental diagnoses of CM type 1 on magnetic resonance imaging (MRI) are symptomatic.

Methods: Our study is a retrospective analysis of the clinical, radiological, and therapeutic data concerning 17 patients admitted to neurosurgery between 2008 and 2018 for Chiari malformation type I.

Results: The mean age of patients included in this study was 39 (extremes ranging from 21 to 58 years). 10 male and 7 female were recorded. Headaches (48%), neck pain (30%), sensory disturbances (26%) and weakness of the 4 limbs (26%) were the most common presenting complaints. An MRI was performed in all cases. A syringomyelia was present in 82.3% of the cases and the most frequent location being cervical. A decompression of the suboccipital bone and posterior arch osteotomy of C1 through a 2-cm midline incision was performed with duraplasty. The mean follow-up period was 18 months, which showed beneficial outcomes in 52.9% of the cases, a stabilization in 41.2% of cases and a worsened outcome with one patient. Syringomyelia symptoms were improved in 60% of the patients.

Conclusion: Chiari malformation type 1 is uncommon. Unlike Chiari malformation type 2 and 3, it can be latent for a long time, described only in adults. However, since the development of magnetic resonance imaging, CM type I can be diagnosed from childhood.

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