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Case Report

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Arnold-Chiari malformation: a case report

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ABSTRACT

Arnold-Chiari or simply Chiari malformation is the name given to a group of deformities of the posterior fossa and hind brain (cerebellum, pons and medulla oblongata). Chiari I is the least severe and is often found incidentally. It is characterized by one or two pointed (not round) cerebellar tonsil sprotruding 5 mm below the foramen magnum, measured by a line drawn from the basion to the opisthion (McRaeLine). Headache is the most common presenting symptom (60-70%) in CIM cases. It is typically localized to the occipital and/or upper cervical region. The pain is usually paroxysmal and of short duration following Valsalva maneuvers such as coughing, sneezing, laughing, straining. Syringomyelia is observed in 30-70% of CIM patients due to disruption of CSF. Dynamics and progressive scoliosis in some of them. Syringomyelia is most commonly found in the cervical region.

Keywords: Arnold-Chiari malformation, headache, syringomyelia

INTRODUCTION

Arnold-Chiari or simply Chiari malformation is the name given to a group of deformities of the posterior fossa and hind brain (cerebellum, pons and medulla oblongata). Problems range from cerebellar tonsillar herniation through the foramen magnum to absence of the cerebellum with or without other associated intracranial or extracranial defects such as hydrocephalus, syrinx, encephalocele or spinal dysraphism.[1-3]

Chiarimal formations are classified according to the irmorphology and severity of anatomical defects, usually by imaging (orautopsy). Chiari I is the least severe and is often found incidentally. It is characterized by one or two pointed (not round) cerebellar tonsil sprotruding 5 mm below the foramen magnum, measured by a line drawn from the basion to the opisthion (McRaeLine). Chiari II consists of a brain stem herniation and an ascending cerebellum in addition to herniated cerebellar tonsils and vermis due to an open distal spinal dysraphism/ myelomeningocele. Chiari III is a herniation of the hind brain (cerebellum with or without brain stem) into a low occipital or high cervical meningoencephalocele. Chiari IV is now considered obsolete.[4] Before it became an outdated diagnosis, it was a more controversial and rare variant showing severe cerebellar hypoplasia similar to primary cerebellar agenesis. Previously, some have stated that myelomeningocele may be present.[5] While others have argued that the presence of myelomeningocele should be classified as a Chiari II with a vanishing cerebellum.[6] In this case report, we wanted to present a case of Arnold Chiari type 1 who presented with headache and weakness in both upper extremities.

CASE

A 33-year-old patient who has been describing occasional blunt pain in the nape of the neck for about 2 years states that the pain increas esespecially when leaning forward or coughing. He states that the pain has become more frequent in the last 3-4 months and is in the form of a feeling of pressure in the back of the head. He also complained of dizziness, imbalance and occasional ting

Ling and numbness in his hands. In the last 1 month, she has noticed a decrease in fine motor skills (such as difficulty in buttoning buttons). The patient stated that he was sometimes unable to urinate completely and had a feeling of tightness and applied to the urology outpatient clinic with these complaints. No pathology was detected in the urologic evaluation and symptomatic treatment was given but the patient did not benefit.

Neurological Examination

Cranial nevre examination: Normal

Eye movements free in both directions

Direct light reflex/indirect light reflex ++/++

Both upper extremities proximal 4/5, distal 3/5 motor strength

Low erextremity motor examination is normal (5/5)

DTR increased in upper extremities

Bilateral Hoffmann +/+

DTR normal in lower extremities, no pathologic reflexes

Romberg test +

Finger-nose test mildly dysmetric

Segmental decrease in pain and heat sensation (especially in C5-C8 dermatomes)

Mild antalgic gait was observed.

Preoperative cervical MRI image of the patient: Syringomyelia continuing from C2 level to T2 level was observed (Figure 1).

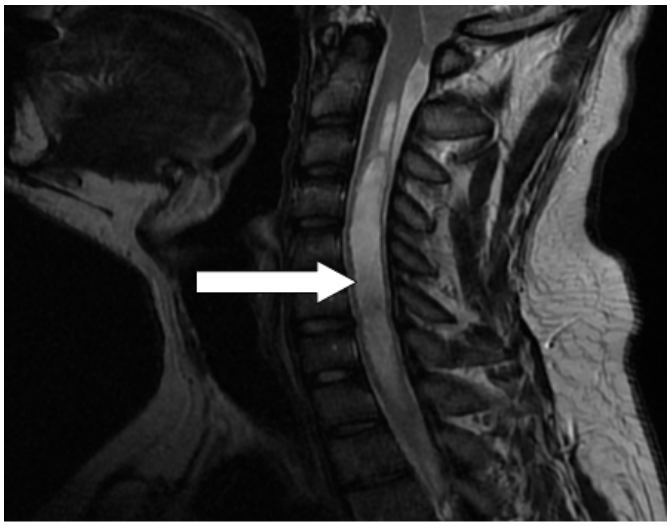

Figure 1. Preop cervical MRI. Sagittal T2 MRI.The patient's cervicothoracally located syrinx is indicated by the whitearrow.
MRI: Magnetic resonance imaging

Postoperative cervical MRI image of the patient: Syringomyelia starting from the C4 level and extending to the C7 level was observed (Figure 2).

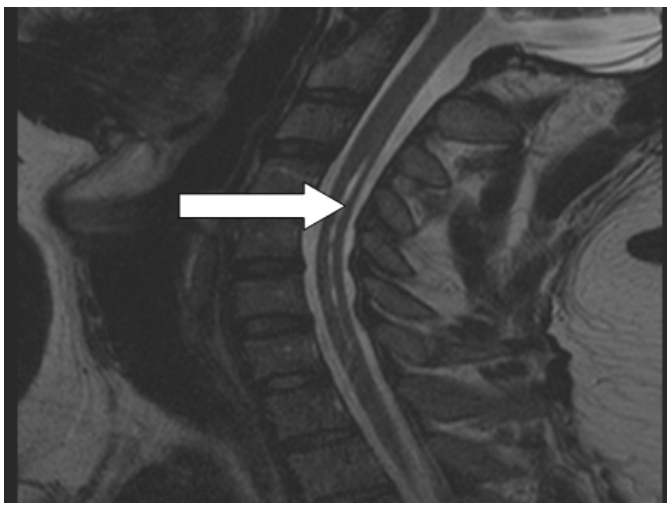

Figure 2. Postoperative cervical MR image Sagittal T2 MR image of the patient's cervicothoracic region with markedly decreased syrinx
MRI: Magnetic resonance imaging

DISCUSSION

Arnold-Chiari or simply Chiari malformation is the name given to a group of deformities of the posterior fossa and hind brain (cerebellum, pons and medulla oblongata). Chiari I is the least severe and is often found incidentally. It is characterized by one or two pointed (not round) cerebellar tonsil sprotruding 5 mm below the foramen magnum, measured by a line drawn from the basion to the opisthion (McRaeLine).[9]

In most cases, the volume of the posterior fossa is small. Syringomyelia in CM1 results from the cerebellar tonsils blocking normal CSF flow through the foramen magnum during the cardiac cycle. Surgical removal of the obstruction to CSF flow results in resolution of the syrinx.[10]

Early observations by Chiari and others suggested a common mechanism of cerebellar ectopia and cerebrospinal fluid disorders (e.g., hydrocephalus and syringomyelia). Critical animal studies by Dorcus Padget advanced our understanding of the embryologic basis of CMI-IV and its relations hiptodysraphism, as in the more severe Chiari malformations and Dandy-Walker malformation.[10] Further studies by Miguel Marín-Padilla supported that these malformations (CMI-IV, Dandy-Walker malformations and various forms of dysraphism) result from various disorders of neuraxial induction.[11]

The most common presentation in Chiari I malformation is suboccipital headaches and/or neck pain (80%). Symptoms are exacerbated when asked to perform the Valsalva maneuver. Other common presentations include eye disorders, auto neurologic symptoms (dizziness, hearing loss, vertigo), gait ataxia and generalized fatigue. Although much less common, the literature reports numerous case studies where patients present with isolated limb pain or weakness, one such report involves the presentation of unilateral shoulder pain with isolated muscle weaknessto a sports medicine clinic.[12]

Myelopathy classically presents with “discrete sensory loss” (loss of pain and temperature sensation, preserved fine touch and proprioception) and motor weakness.[13,14]

Cerebellar findings such as ataxia, dysmetria and nystagmus and lower cranial nevre deficits (IX, X, XI, XII CN) are caused either by direct compression of the cerebellum or medulla in the foramen magnum or by syringomyelia or syringobulnia.

Sleep apnea may occur in a patient with Chiari malformation due to weakness in the pharyngeal muscles caused by compression of the brainstem, upper spinal cordor lower cranial nerve.[15]

CONLUSION

We need to exclude some conditions that come with a clinical presentation similar to Arnold Chiari-like. Intracranial hypotension –may mimic midbrain prolapse, cerebellar tonsilor posterior brain herniation. Normal variant cerebellar tonsil ectopia -does not meet the criteria for Chiari malformation and is an incidental finding in an asymptomatic patient. Cerebellar tonsillar herniation caused

by increased intracranial pressure (ICP)-ICP causes such as neoplasm, hydrocephalus, mass effect from trauma or hemorrhage should be evaluated.

With early diagnosis and close follow-up in Arnold chiari malformation, we can make the correct diagnosis and maximize the patient's life standard with surgical operation at the most appropriate time.

ETHICAL DECLARATIONS

InformedConsent

The patient signed and free and informed consent form.

Referee Evaluation Process

Externally peer-reviewed.

Conflict of Interest Statement

The authors have no conflicts of interest to declare.

Financial Disclosure

The authors declared that this study has received no financial support.

Author Contributions

All of the authors declare that they have all participated in the design, execution, and analysis of the paper, and that they have approved the final version.

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