ACTA MEDICA SALINIANA

ISSN 0350-364X

Acta Med Sal 2009; 38 (1): 44-46

CASE REPORT

ARNOLD–CHIARI MALFORMATION AND SYRINGOMYELIA

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Received: 03.04.2008
Accepted: 09.09.2008

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ABSTRACT

Generally, Arnold – Chiari malformation associated with syringomyelia is not rare. In this case report we present a 52 years old female patient with a history of neck-pain, low-back pain, pain in both arms as well as frequent numbness in the lateral regions of both arms. She also experienced walk disturbance and in her previous history she reports a car accident, twenty-eight years ago. She broke windshield with her forehead, but remained conscious at the time. At the time of admittance at the Department of Neurology, her neurological status was remarkable for cerebellar symptomatology (ataxia, positive Romberg-sign, «finger–nose» test which she did with tremor and was not able to perform «tandem-walk»), hypoestesia of the right side of the body, hyper-reflection of both arms and legs, bilaterally, more impressive to the right; right foot subclonus and spastic-ataxic walk. Neurological status could not be explained by previously performed tests: x-ray of cervical spine, lumbal myelography, computerized tomography (CT) of the brain. However, magnetic resonance imaging (MRI) of the brain and cervical spine verified Arnold-Chiari malformation (type I) associated with syringomyelia from C3 to C7. Patient was referred to surgery and subsequent physical rehabilitation.

Keywords: *Arnold – Chiari malformation, syringomielia*

INTRODUCTION

The Chiari I malformation (CMI) is a caudal displacement of the cerebellar tonsils into the cervical spinal canal. It is generally agreed that CMI is defined by tonsillar herniation more than 5 mm below the plane of the foramen magnum. The disorder affects children and adults and may be congenital or acquired. It is originally described by Arnold in 1894 and Chiari in 1896.[1,2] The exact cause of the Chiari malformation is unknown. It has been suggested that during early embryo development of the brainstem and spinal cord, the malformation occurs. The incidence of CMI is not known. Before the availability of nuclear magnetic resonance imaging (MRI), CMI rarely was diagnosed. Recently, an incidence of 0.6% was reported in all age groups, and an incidence of 0.9% was reported in a study of only pediatric patients.[3] In patients with CMI, the most common presenting symptom is pain, but also some presenting signs include brainstem, cerebellar, and spinal cord dysfunction: oculomotor (17.1%), vestibulocerebellar (84.8%), bulbar (35.4%), conduction motor (25.9%) and segmental motor sensory disturbances (9.5%).[4] Although advances in MRI have significantly enhanced our ability to diagnose CMI, management of this condition remains controversial. Syringomyelia is a chronic progressive degenerative

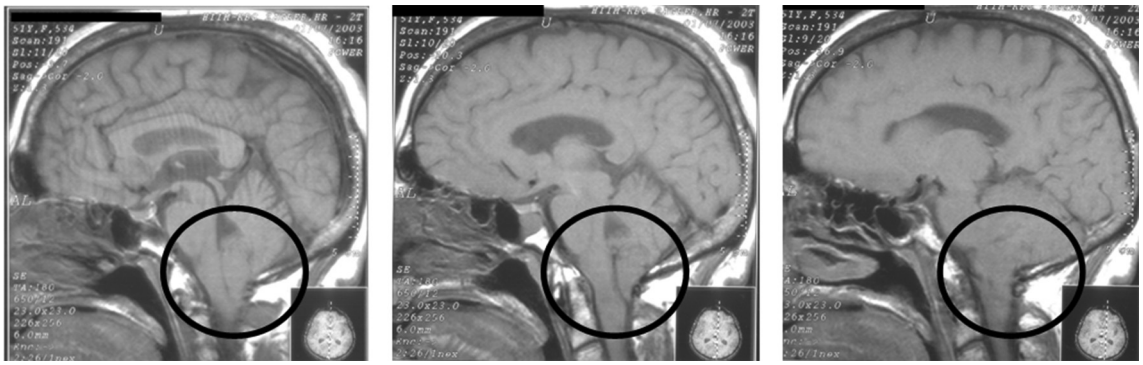

Figure 1. *Arnold-Chiari malformation (type I) in 52 years old female patient visualized by magnetic resonance imaging of the brain*

disorder of the spinal cord, characterized clinically by brachial amyotrophy and segmental sensory loss of dissociated type, and pathologicaly by cavitation of the central parts of the spinal cord, usually in the cervical region but extending upward in some cases into medulla oblongata and pons (syringobulbia) or downward into the thoracic or even lumbar segments.[5] Syringomyelia associated with CMI is commonly between the C-4 and C-6 levels.[6]

Although the exact development of syringomyelia is unknown, there are many theories about the formation of a syrinx cavity. One theory suggests that the herniated tonsils (Chiari I Malformation) result in a plug at the outlet of CSF from the brain to the spinal canal. There are various data concerning presented both CMI and syringomyelia.

Distention with cerebrospinal fluid (CSF) of the central canal of spinal cord (hydromyelia) or paracentral cavities (i.e., syringomyelia) is present in approximately 25% of patients with CMI3 to 40%.[6]

CASE REPORT

We present a 52 years old female admitted at the Department for Neurology of the University Clinical Center Tuzla with a history of neck-pain, low-back pain, pain in both arms as well as frequent numbness in exterior parts of both arms. Also, she experienced walk disturbance. She has been suffering from these symptoms for several years. Neurologists as well as physical medicine specialists have examined her several times. Twenty-eight years ago she had a car accident. She broke windshield with her forehead, but remained conscious. At the addmission at the Department of Neurology, her neurological status was remarkable for cerebellar symptomatology (ataxia, positive Rombergsign, «finger–nose» test which she did with tremor and was not able to perform «tandem-walk»), hypoestesia of the right side of the body, hyper-reflection of both arms and legs, bilaterally, more pronounced at the right side; right foot subclonus and spastic-ataxic walk.

Before she has been admitted, several neurodiagnostic procedures have been performed: x-ray of the cervical spine showed disc-arthrosis C4 to C7 and spondylosis C4 to C7, x-ray of the lumbo-sacral segment depicted degenerative spondylosis and computerized tomography (CT) of the brain was normal. Auditory and visual evoked potentials were normal. Results of these procedures could not explain her neurological status. After MRI of the brain and cervical spine we verified CMI (Figure 1), associated with syringomielia from C3 to C7 levels (Figure 2, Figure 3).

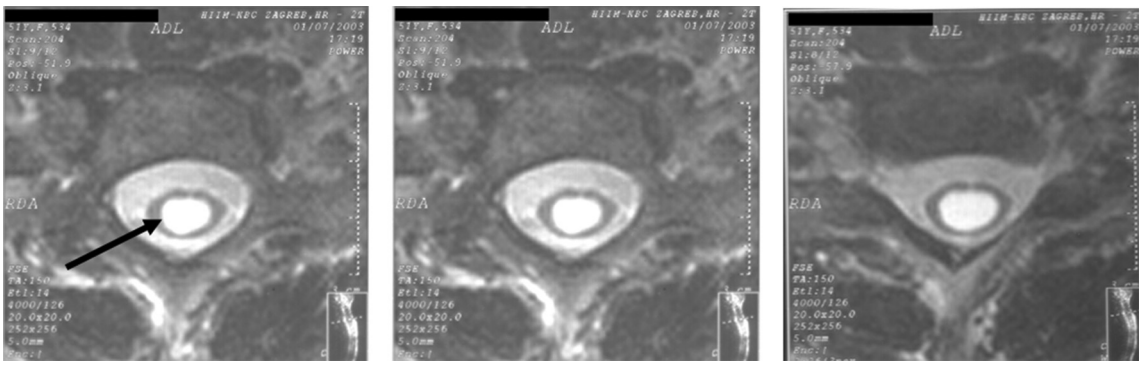

Figure 2. *Syringomyelia*

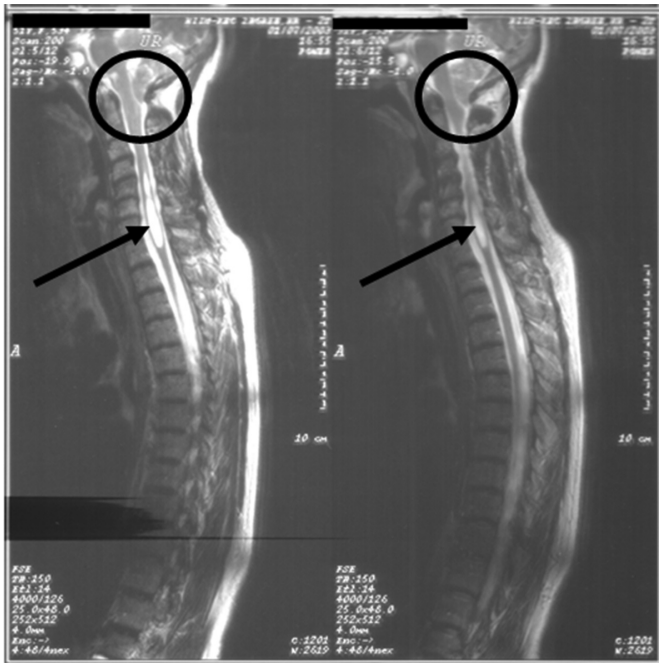

Figure 3. *Both Arnold-Chiari malformation (see circle) and syringomyelia (arrow)*

DISCUSSION

Patients with CMI remain asymptomatic for a prolonged periods of time. Arachnoidal scarring and adhesions are suggested to build up at the foramen magnum as the cerebellar tonsils rub against bone over the years. Arachnoidal adhesions may increase the compression of the hindbrain and spinal cord and further interfere with CSF flow at the foramen magnum and thus cause symptoms. These adhesions are often discovered at the time of surgery; however, many patients remain symptomatic despite adequate surgical decompression of the foramen magnum. Many patients report trauma, such as whipsplash injuries and direct blows to the head and neck, as the precipitating event for symptom onset. Such trauma may accentuate tonsillar impaction or result in a subarachnoid hemorrhage that destabilizes a marginally compensated CSF system.[3] The onset of symptoms may be delayed during life. There may be signs and symptoms of injury to the cerebellum, medulla, and the lower cranial nerves, with or without evidence of increased intracranial pressure. Progressive ataxia, leg weakness, and visual complaints are characteristic.

The signs and symptoms of the Arnold-Chiari malformation in adults may simulate the syndromes produced by tumors of the posterior fossa, multiple sclerosis, syringomyelia, or basilar impression. Once these "onset of symptoms" occurs, the most frequent treatment is decompression surgery, in which a neurosurgeon seeks to open the base of the skull and re-establish CSF flow to the spine. Decompression is a very taxing surgical procedure and is now, in some circles, disdained in lieu of tethered cord detachement at the base of the spine. Some surgeons find that detethering the spinal cord relieves the compression of the brain against the skull opening (foramen magnum) obviating the need for decompression surgery and associated trauma. It should be noted that the alternative spinal surgery is not without risk.[7] Our patient experienced head trauma many years before symptoms appeared. After verification of CMI, the patient was referred to surgery and subsequent physical rehabilitation.

A month and a half after the physical rehabilitation the patient was well recovered, until she started re-experiencing same symptoms as before the surgery: neck pain and walking disturbances.

CONCLUSION

In our clinical practice Arnold-Chiari malformation itself, as well as associated complications (syringomyelia), are not often. Negative results of standard neurodiagnostic tests in diagnostic work-up of chronic symptoms in patients with a columnar disorder, especially craniocervical junction, may be misleading for a neurologists, Therefore, it may be necessary to perform MRI of the brain in order to definitely determine the ethiological cause of symptoms.

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