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Untreated Chiari 1 Malformation in Adulthood with Massive Hydrosyringomyelia and Hydrocephalus

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Abstract

We describe a case of untreated Chiari I malformation in adulthood. It is characterized by displacement of the cerebellar tonsils caudally through the foramen magnum, associated with massive hydrosyringomyelia and tetraventricular hydrocephalum. Magnetic Resonance (MR) imaging provides excellent visualization of intracranial and intraspinal malformations, and thus contributes greatly to their correct diagnosis and treatment.

Keywords

Chiari 1 malformation, Tetraventricular hydrocephalum, Hydrosyringomyelia, Adulthood, Magnetic resonance, Diagnosis

Introduction

Chiari I malformation is the most common variant of the Chiari malformations. It is a dynamic disease characterized by displacement of the cerebellar tonsils caudally through the foramen magnum, interfering with the physiological flow of cerebrospinal fluid [1]. Although is still considered a rare condition, has been recently reported a prevalence rates of 0.1-0.5% with a slight female predominance [2].

Often, it remains asymptomatic until adulthood. The likelihood of becoming symptomatic is proportional to the degree of downward descent of the tonsils.

The most common symptoms include headache and neck pain often exacerbated by cough and Valsalva maneuver. Hydrocephalus and Syringomyelia occurs less frequently and are usually associated with asymmetrical central cord symptoms such as hand weakness and dissociated sensory loss [3].

Magnetic Resonance (MR) is considered the imaging modality of choice [4]. The diagnosis is made by measuring how far the tonsils protrude below the margins of the foramen magnum; a displacement of tonsils > 6 mm from the inner margin of foramen magnum is considered diagnostic for Chiari I malformation [5]. MR imaging provides also excellent visualization of intracranial and intraspinal complications, addressing the proper diagnosis and treatment planning.

Treatment of Chiari malformation depends on the severity and associated symptoms. Regular monitoring, medications and surgery are treatment options. Surgery is usually reserved only for patients with syrinx and symptoms [6]. It consists of decompressing the posterior fossa, by removing part of the occipital bone, and posterior arch of C1.

Despite is not a common condition, it is mandatory to make a prompt and correct diagnosis as soon as possible. Unrecognised Chiari I malformation can leads to severe symptoms and massive complications, as reported in our case. We describe a unique case of untreated Chiari 1 malformation in adulthood associated with massive hydrosyringomyelia and tetraventricular hydrocephalus.

Case Report

A 35-year-old girl was admitted to our hospital because of progressively increasing problems with walking for five years. She had a severe scoliosis, she occasionally complained achy joints and she was showing apathetic behaviour. Neurological examination revealed normal mental functions, Bernard Horner syndrome to the right (Figure 1), spastic gait, positive Romberg, diminished motor strength in her legs and right arm with muscle atrophy in the same hand (4 out of 5) (Figure 2, claw hand). Pain and temperature sensation was impaired in both arms, in her face; chest and abdomen and vibration sense was lost in lower extremities while other sensations were preserved. There were brisk reflexes but in her right arm where they were only slightly diminished (1+), clonus in her left ankle, left positive Hoffmann sign and bilateral Babinski's reflex. She complained nocturnal incontinence. Cerebellar tests including

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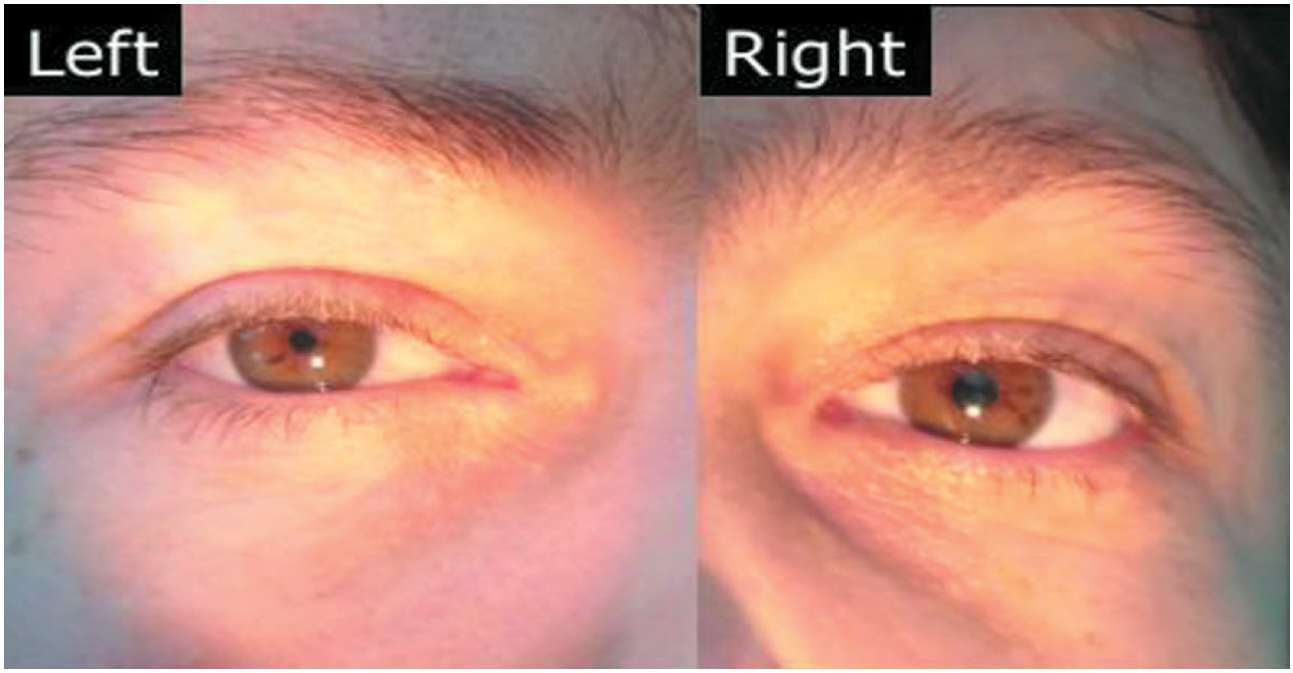

Figure 1: It's clearly visible a Bernard Horner syndrome on the right eye, characterized by constriction of the pupil (miosis) and mild dropping of the eyelid (ptosis).

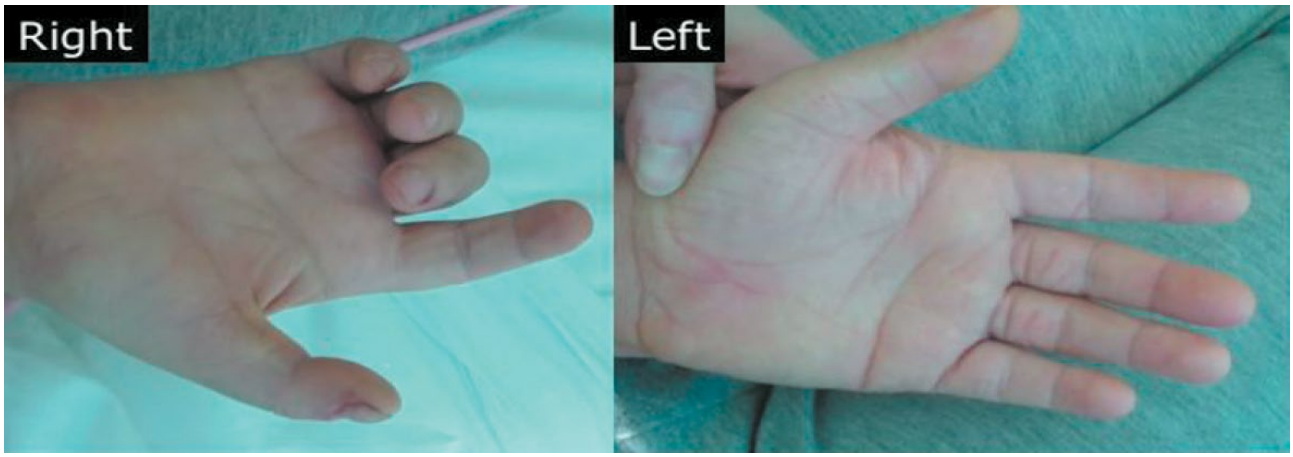

Figure 2: In this figure, on the right, is reported a claw hand, a deformity manifested by flattening of the transverse metacarpal arch and longitudinal arches, with hyperextension of metacarpophalangeal joints and flexion of the proximal a distal interphalangeal joints.

finger to nose, heel to shin test and rapid alternating movements were normal.

A spinal and cerebral MR examination was performed showing an extensive cylindrical hydro syringomyelia extending from C1 to L5 (Figure 3, Panel A). Complete medullary atrophy was observed with only residual fascicula identified (Panel B). No spinal dysraphisms were present. Downward displacement of the cerebellar tonsil at C2 level was noticed (Panel C) with associated horizontally oriented short clivus and posterior angulation of the odontoid process (Panel A, *arrow*), causing brain steam compression with flow void in the ependymal canal due to increase liquoral velocity. A severe tetra ventricular hydrocephalus with marked atrophy of the cerebral parenchyma was also present (Panel D). Patient was referred to surgical decompression.

Discussion

Our case perfectly illustrates the pathophysiological evolution and features of untreated Chiari 1 malformation.

The underestimation of early symptoms and the lack of a proper imaging evaluation as first step, can lead to massive life-threatening complications. Longstanding brain steam compression causes an increased cerebrospinal fluid (CSF) pressure with syrinx and hydrocephalus formation. Patients with Chiari 1 develop these anomalies respectively in 30-70% and 10% of cases [1,7,8]. Other foramen magnum or spinal pathologies that impede CSF flow can determine syrinx formation, including post-infectious arachnoiditis, spinal cord/column injury, cervical stenosis and basilar impression [9].

In the setting of Chiari malformation, cervical tether release or foramen magnum decompression represent effective interventions targeting the syrinx mechanism. The reestablishment of normal CSF flow has been demonstrated to allow syrinx resolution or stabilization; otherwise symptoms can progress to severe pain or disabling neurologic impairment [10], as reported in our case. MR represents the reference imaging technique in Chiari assessment and preoperative planning, evaluating the volume of the posterior fossa, CSF flow dynamics, medullary anomalies and spinal dysraphism. Computed Tomography can add useful information in case of complex bone anomalies. In conclusion, we reported a rare case of untreated Chiari I malformation in which the underestimation of early symptoms leaded to massive hydrosyringomyelia, hydrocephalus and to the development of serious neurological impairment.

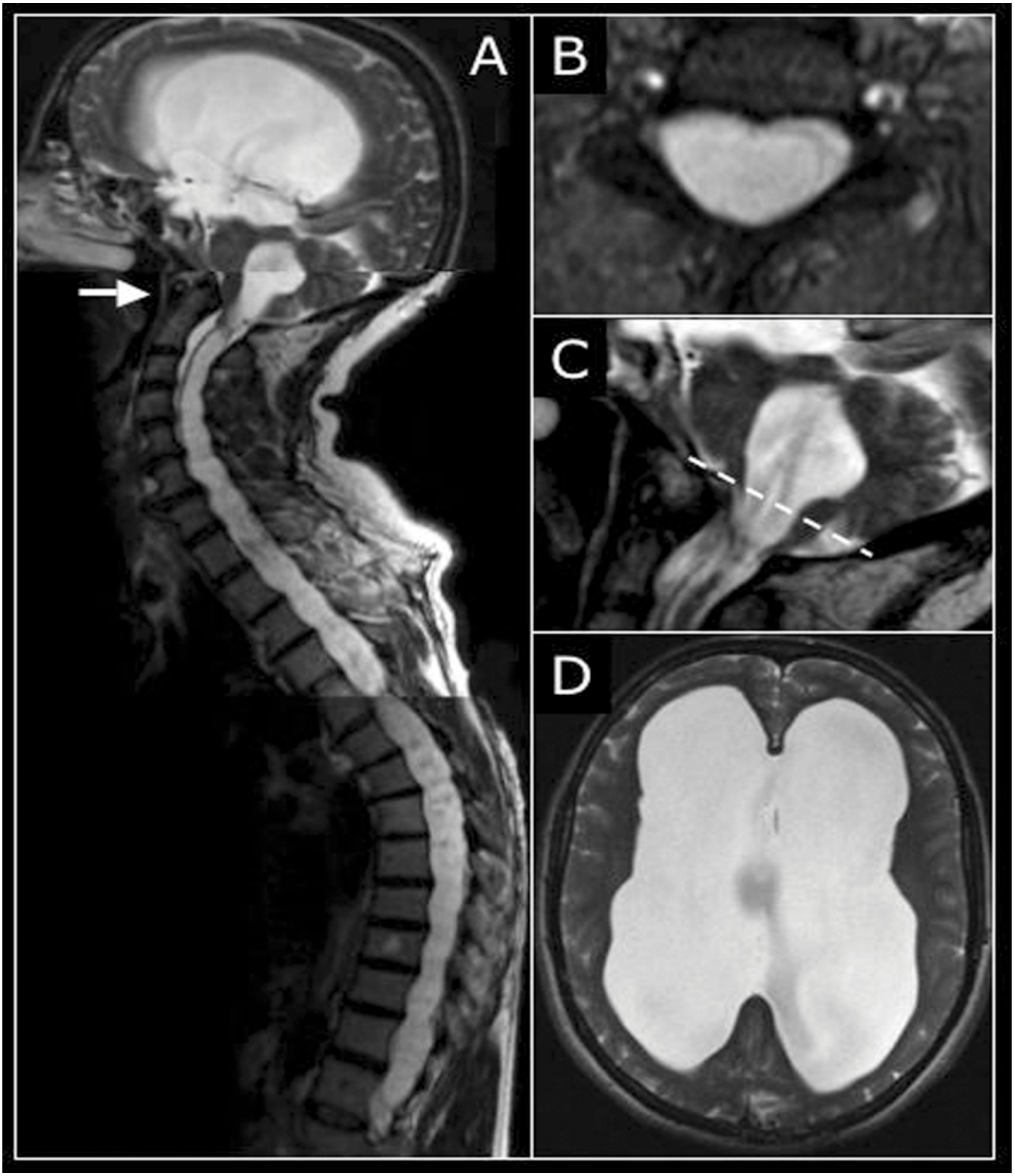

Figure 3: This figure reported a spinal and cerebral MR examination, showing key findings of Chiari 1 Malformation with Massive Hydrosyringomyelia and Hydrocephalus.

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