Cruciate hemiplegia associated with basilar invagination, chiari malformation and syringomyelia in children: Case report and review of literature

Hemiplejia cruzada asociada con invaginación basilar, malformación de chiari y siringomielia en niños: Reporte de caso y Revisión de la literatura

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Resumen

Presentamos un caso de hemiplejía cruzada en un paciente de 12 años que presentó síntomas progresivos de parálisis simultánea del brazo derecho y de la pierna izquierda. La investigación radiológica mostró invaginación basilar (BI), malformación de Chiari (CM) y siringomielia. El paciente fue operado a través de una craneectomía de la fosa posterior y duroplastia. Seguimiento con buen resultado y mejora significativa de la marcha.

Palabras clave: Hemiplejia cruzada, invaginación basilar, malformación de Chiari, siringomielia.

Abstract

We report a case of cruciate hemiplegia in a 12 year-old patient who presented progressive symptoms of simultaneous right arm and left leg paralyses. Radiological investigation showed basilar invagination (BI), Chiari malformation (CM) and syringomyelia. The patient was operated through a posterior fossa craniectomy and duroplasty. Follow up with good outcome and significant gait improvement.

Key words: Cruciate hemiplegia, basilar invagination, Chiari malformation, syringomyelia.

Introduction

Cruciate hemiplegia is characterized by paralysis of upper limb and contralateral lower limb resulting from injury at the decussation of the medullary pyramids[17,19]. Some etiologies related to this rare syndrome have been reported in adults, such as ischemic events, hemorrhages, traumas and craniocervical malformations, including basilar impression (BI), Chiari malformation (CM) and syringomyelia[2,3,9,10,15,19].
We report a case of cruciate hemiplegia in a 12 year-old patient associated with the BI, MC and syringomyelia that was operated and had clinical improvement. To our knowledge, this is the first reported case from Brazilian pediatric population.

Background and clinical presentation

Cruciate paralysis is a rare incomplete medullary syndrome. Bell, in 1970, was the first to describe cruciate paralysis as a clinical entity, when he published a series of four cases[1]. Unlike the central cord syndrome described by Schneider[15], we can find involvement of lower cranial nerves. The most affected nerve is the accessory nerve, and there can be multiple cranial nerve palsy. Urinary retention and respiratory dysfunction are also described[4].
After review of the literature (PubMed, SciELO, Cochrane), the cruciate paralysis in children was described only in a head trauma case, with C1 fracture and paralysis of upper limbs associated to Chiari malformation on MRI, this feature being considered an aggravating factor[5]. A rare clinical manifestation of involvement of a member and the other contralateral, similar to our case report, was described in 1996 in a Brazilian woman of 56 years with similar symptoms and also with craniocervical junction malformation (BI, CM and syringomyelia), without trauma[9].
Our patient had motor deficit, superficial and deep sensory deficit, and cerebellar syndrome. It could not be explained only by a focal injury at the medulla. The motor and sensory disturbances were possibly caused by bilateral lesions involving the corticospinal fibers in the pyramids, lemniscus, and lateral spinothalamic tract, at the ventromedial and dorsolateral regions of the medulla, probably induced by craniocervical junction malformations (BI, CM and syringomyelia).
The functional and anatomic characteristics of the cruciate paralysis are complex. Some authors have described motor pathways injuries at the decussation of the medullary pyramids, with consequent quadriparesis or quadriplegia[9]. The medullary injury can happen as complication of surgical procedure for craniocervical malformations, for example, compression of the bulbar pyramids by the odontoid process, aggravated by bending the head during surgery[7,8]. Other studies report as a result of cervical spine trauma associated with C1, C2 and C3 fractures, firearm projectile injuries, rheumatoid arthritis, infections and tumors[2,5,6,12,16].
Anatomic aspects of the pyramidal decussation at the medulla can try to explain the crossed type of the motor deficit. According to Wallenberg (1901), the pyramidal fibers to the upper limbs cross more cranially than the fibers to the lower limbs. In contrast, Oppenheim (1923) proposes that the pyramidal fibers to the lower limbs would cross higher than those to the upper limbs[1,13,14].

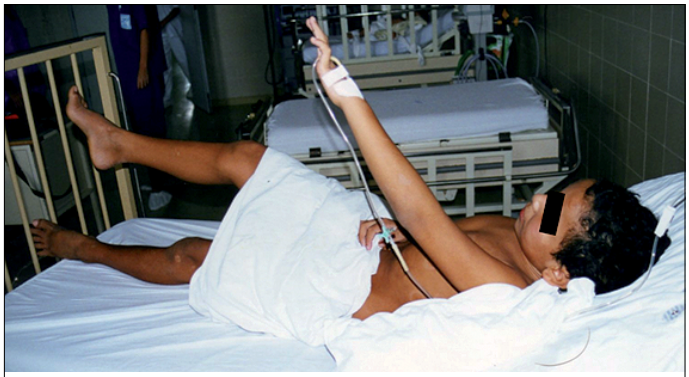

Figure 1. Severe right arm and left leg paresis (Mingazzine maneuver).

Diagnosis

A differential diagnosis reported in the literature is the "corticarlis bimembris monoplegia" (false cruciate hemiplegia), due to bilateral ischemic lesions on two different territories, as an injury affecting the territory of the anterior cerebral artery, and the other in the territory of the middle cerebral artery in the contralateral hemisphere, with leg and arm monoplegia respectively[14].
MRI is the gold standard for encephalic lesions, especially at the brainstem. It is essential to diagnosis of underlying disease causing the cruciate paralysis. If present, it can characterize craniocervical junction malformation as well[4,11].

Management and outcome

The treatment is individualized depending on the underlying disease, but usually with good outcome[11].

Case description

A 12 year-old boy presenting with progressive gait disturbance and left leg and right arm numbness for 6 months. A month before hospital admission he complained of severe weakness on left leg, gait worsening (could not walk), and nuchal pain. Neurological examination showed "brevis collis", low-set hair, vertical nystagmus, severe right arm and left leg paresis (Figure 1), with hypertonia and asymmetrical hyperreflexia (4/4 + member in the upper right and left lower limb; 3/4 + in the left upper limb and lower right). Left Babinski sign and left clonus were present. Syringomyelic dissociation in the left arm and chest, and hipopalestesia of four limbs were also present. Magnetic resonance imaging (MRI) (Figure 2) showed basilar impression (BI), Chiari malformation (CM) and syringomyelia.
He underwent posterior fossa decompression through an occipital craniectomy and C1 laminectomy. After opening the dura, herniation of the cerebellar

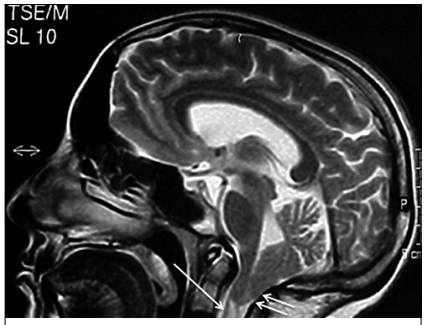

Figure 2. MRI, T2 sagittal. Basilar impression, Chiari malformation (tonsillar herniation, double arrow), and cervical syringomyelia (spinal cord hyperintensity, long arrow)

tonsils until the beginning of C2 was evidenced. Intra-pial tonsil aspiration was performed as recommended (Williams[18]) and laterally attached to the dura. The fourth ventricle was blocked, and decompression was performed followed by duroplasty.

The patient had good outcome, with significant gait improvement. He started walking with crutches after 3 months, and without medical device after 12 months.

Conclusion

Cruciate paralysis can be associated with craniocervical junction malformations, resulting in arm paralysis alone, although there may be involvement of the lower limbs. This syndrome should be kept in mind in children with basilar impression, Chiari malformation and/or syringomyelia, with or without head trauma.

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