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Siringomielia, Report of A Clinical Case and Bibliographic Review

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1. Summary

1.1. Introduction: Syringomyelia is a pathological affectation of the spinal cord, which consists of a central cavitation, which mainly affects the cervical area. In 90% of cases, it may be closely related to Chiari's disease, however, there are other alterations such as intra or extramedullary tumor, trauma, hydrocephalus, among others that can cause syringomyelia as well. Diagnosis is completed with imaging (MRI) studies. Treatment is usually surgical in most cases if the patient is a candidate for it. However, there are other alternatives with promising results.

1.2. Clinical Case: Female patient 47 years of age, who attends the external consultation referring pain in the upper extremities mainly on the right, radiated to dorsal region, in addition to presenting other signs and symptoms such as tachycardia, medium intensity precordial pain, headache, nausea and dizziness. As a pathological personal record, the patient refers to having suffered seizures during her childhood, as well as being hit by a truck with an evolution time of 15 years.

1.3. Conclusions: with the studies obtained and the evaluation of the patient, the diagnosis of syringomyelia associated with Chiari is reached. The patient is tracked and scheduled for neurosurgical treatment due to the presence of symptomatology.

2. Introduction

Syringomyelia is a generic term for a disorder in which a cyst or tubular cavity forms inside the spinal cord. This cyst is known as syringe or "syrinx". It must be differentiated from the term hydromielia which refers to a dilation of the central medullary canal [1].

It is a unique pathological entity of the spinal cord that consists of a generally central cavitation and that preferably affects the cervical marrow. This pathology is progressive and degenerative presenting fluctuating tables ranging from severe worsening in 5 to 10 years to a slow progression of 30 to 40 years or more [3].

In 90% of cases, syringomyelia is associated with Chiari type 1 malformation, while the remaining 10% include unions association with Chiari1 (idiopathic), which may be secondary to intramedullary tumor, spinal trauma, extramedullary tumor compression, spinal infarction, and hydrocephalus [7].

Chiari's disease is characterized by being an anatomical abnormality that causes the lower cerebellum (cerebellar tonsils) to move out of its normal position at the back of the head to the cervical (or neck) region of the spinal canal. In the case of trauma or tumor complication, syrinx, or cyst forms in a segment of the spinal cord, which begins to expand, subsequently causing symptoms, with pain being the main pain. If the involvement involves the brain stem (syringomyelia), alterations in vital functions such as breathing and heart rate may occur [2, 3, 5].

The clinical picture usually begins in the third decade of life. At first it can manifest itself unfinished with pain in the posterior cervical area, being able to be triggered by physical exertions or maneuvers of Valsalva and subsequently present hypoesthesally mainly in upper limbs, as well as motor deficit in them, with minimal involvement in the lower limbs [5].

Diagnosis is based on good anamnesis as well as a complete physical examination, emphasizing neurological functions. Subsequently, imaging studies can be requested, with MRI being the study of choice in these cases, as it allows to see in detail the structures to be studied, as well as the alterations that may occur, mainly in the cerebrospinal fluid and spinal cord [7].

Treatment in patients with this pathology is usually highly variable; Surgery is generally recommended, with the aim of providing more space for the cerebellum (Chiari malformation) at the base of the skull and upper neck, without entering the brain or spinal cord. This allows the primary cavity to flatten or disappear and thus the symptomatology improves [2, 4].

Drug treatment has no curative value, radiation is used infrequently and produces few benefits. In the absence of symptoms, you are generally not given any treatment [6].

There should be a protocol with which candidate patients can be chosen for surgery, these should have significant symptomatology such as severe chronic pain, limitation in daily activities, respiratory problems, persistent seizures, adequate surgical risk by internal medicine, denied commodities or under treatment. All this to always assess the risk-benefit of the patient [6].

3. Clinical Case

Female patient, of name referred to by the following acronym MCGD of 47 years of age, who enters the emergency room for a picture of seizures, presented at 03:00hrs at home, approximately three minutes long with loss of consciousness, referred to by her daughter who sensed the act; mild frontal concussion. The patient refers to having the history of seizures in childhood, without presenting from the age of 19. Already in the emergency department, physical examination is consenting, oriented, without neurological deficit, at that time stable; vital signs within normal parameters, isoreflective pupils, cylindrical neck without palpable adenomegalys, cardiac area without added noises, well-ventilated pulmonary fields, globose abdomen at the expense of adipose, depressible panicle, no pain to superficial or deep palpation, upper extremities with decreased strength, increased osteotendinous reflexes, hair filling of 2 seconds, rest of the physical examination without alterations. You are diagnosed with "seizures, not classified elsewhere". As well as its respective handling with hydration, safety position and antiepileptic, it is kept under observation, and subsequently discharged from the emergency department by appointment at the external consultation for the neurology service.

He then goes to his consultation for the neurology service of the Military Hospital of Medical Specialties of Guadalajara, for referring pain and alteration of force in both upper extremities. The patient refers that the pain is most severe in the upper right limb and in cervical-dorsal region. In addition to tachycardia, medium intensity precordial pain, headache, nausea, and dizziness, with low response to analgesic treatment.

To interrogation, the patient refers to the antecedent of having been hit by a truck of 15 years of evolution, currently with painful sequels in upper right limb and lateral region of the ipsilateral neck. You are given an imaging study (magnetic resonance imaging), which shows data on syringomyelia and cervical osteoarthritis (Figure 1). MAGNETIC resonance imaging, sagittal cut, with suggestive data of osteodegenerative disc disease, with loss of height of the vertebral bodies of C5, C6 and C7; as well as dehydration of the intervetebral discs C5-C6, C6-C7 and C7-T1. In addition to hyperintte dense intramedular cystic cavity in T2 weight of MRI, from intramedullary C4-T2 level. Presenting descent of cerebellar tonsils that obstruct the foramen magician more than 5mm. (Courtesy of the Regional Military Hospital of Medical Specialties of Guadalajara)

The patient was asked to interconsult with the neurosurgery service, to see the possibility of undergoing some surgical procedure due to chronic cervicalgia. He continues to attend consultation with the NEurology service for its management and evolution, as well as the rehabilitation service. It is currently kept waiting for surgical time.



Figure: MAGNETIC Resonance Imaging

4. Discussion

In this case, it is suspected as the first possibility that syringomyelia may be secondary to or related to Chiari's disease. The plan will be to propose neurosurgeon management, prior to which you should have a 4-limb electromyography to corroborate the etiology of neurological damage. Electromyography is a study that will measure the rate of conduction of nerve transmission to infer the etiological origin of the damage if it exists in this patient [8].

This relationship with Chiari's disease, is coupled with the decline

of cerebellar tonsils, which allow us to suspect secondary damage to the disease of the patient. In cabinet studies conducted by the seizure, an intramedullary hyperinte dense image from C4 to T3, suggestive of syringomyelia, was evident as an incidental finding. It should be noted that this injury is already causing significant neurological damage in our patient, however, the diagnosis was obtained through an incidental finding.

5. Conclusions

It is recommended to expand the study and knowledge of this common but underdiagnosed pathology, to provide more and better treatments to patients with this disease.

This will prevent the progression of the disease and promote an early recovery that generates an increase in the quality of life in this type of patients, since it can become disabling, thereby increasing alterations in its biopsychosocial environment.

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