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Research Article

MAGNETIC RESONANCE: UTILITY IN THE DIAGNOSIS OF SECONDARY ENCLOSURE SYNDROME TO MYELINOLYSIS PONTINE

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Abstract

The cloistered syndrome (SE) is a rare disease secondary to osmotic demyelination, which occurs clinically in patients with a progressive alteration of their motor capacity, which severely affects the quality of life of this type of patient and in the long term leads to death, due to the total loss of basic functions. In the following review of cases, the usefulness of magnetic resonance imaging (MRI) as a diagnostic method of this pathology is analyzed and a brief comparison of its effectiveness with computed tomography (CT) is made, which in turn constitutes a highly used imaging test in recent years for the diagnosis of it.

Keywords: Cloistered syndrome, Magnetic resonance imaging, Central pontine myelinolysis, Diagnosis.

INTRODUCTION

Cloistered syndrome (LIS) is a complex medical condition that presents with quadriplegia, bulbar palsy, and sensory loss of the entire body due to damage to the brainstem, most commonly the anterior pons. In those patients with this condition, cognition, vertical eye movement, blinking, and hearing are preserved (1). Patients present quadriplegia due to the involvement of the corticospinal tracts, the gaze is not affected due to the location of the nucleus in the rostral portion of the midbrain (2). Medial and lateral gaze palsies are common, as are diplopia and blurred vision (3). Respiration is often affected in those patients in whom the lateral segment is involved (4). These patients can present different breathing patterns such as Cheyne-Stokes, apneustic and ataxic (Table 1). The LIS has subtypes: Classic, Partial, and Total LIS. Classic LIS has characteristics such as loss of movement in all four limbs and anarthria. In partial LIS, the patient still has preserved motor function. The complete LIS has the worst result since, in addition to quadriplegia, they cannot blink or have vertical gaze (5). The locked-in syndrome is caused by any injury that affects the ventral pons and midbrain; including vascular lesions, masses, infections and demyelinating disorders, especially due to central pontine myelinolysis. Central pontine myelinolysis (CPM) is a demyelinating disease of the pons often associated with demyelination of other areas of the central nervous system (CNS). Which classically occurs in alcoholics, malnourished individuals, chronic liver diseases and rapid correction of metabolic disorders such as hyponatremia (6).

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In the acute setting, CT or MRI of the brain helps to characterize the causative agent to establish a treatment earlier. (7) Imaging studies are the most valuable modality used in the diagnosis of cloistered syndrome. The addition of a CT or MRI angiogram can show vascular lesions causing the syndrome in cases of stroke or arterial dissection. The addition of contrast medium can delineate masses such as tumors or abscesses and even active demyelinating lesions (8). Magnetic resonance is the main method of diagnosis and is superior to computed tomography.

METHODOLOGY

In this review article, a detailed bibliographic search of information published since 2014 was carried out, in the databases PubMed, Elsevier, Scielo, national and international libraries. We used the following descriptors: cloistered syndrome, magnetic resonance imaging, central pontine myelinolysis, diagnosis. The data obtained ranges from 2 to 15 records after using the keywords. The search for articles was carried out in Spanish and English, limited by year of publication, and studies published since 2014 were used.

RESULTS

Min Kyun et al in their case report of a 44-year-old Korean man admitted to hospital due to a dizziness-induced fall and a 1-minute loss of consciousness. The patient had a history of drinking two or more bottles of alcohol every day and had not eaten in the previous month. On initial physical examination, he was alert and no focal neurologic abnormalities were noted.

Table 1. Clinical features of locked-in syndrome

Clinical features		
Quadriplegia	Apneustic respiratory pattern	Preserved consciousness
Anarthria	Emotional liability	Preserved vertical gaze and blinking
Normal o absent sensation	Vértigo	lateral gaze palsy
Cheyne-Stokes respiratory pattern	Insomnia	Preserved hearing
Ataxic respiratory pattern	Internuclear ophthalmoplegia	Impaired attention and memory (early stages)

Laboratory data revealed hyponatremia (serum sodium 118 mEq / L), hypokalemia (potassium 2.5 mEq / L), hypoalbuminemia (albumin 3.2 g / dL), abnormal liver function (aspartate transaminase 175 IU / L and alanine transaminase 68 IU / L). The serum sodium concentration was corrected to 134 mEq / L during the first 14 hours and 137 mEq / L within 18 hours after intravenous injection of 3% saline. One week after admission, he developed progressive dysarthria, dysphagia, and limb weakness, for which he was transferred to the intensive care unit. Nasogastric and tracheostomy feeding tubes were inserted and ventilator care began. Brain magnetic resonance imaging (MRI) showed symmetric areas of signal hyperintensity in the bilateral pons on T2-weighted and diffusion-weighted images, consistent with a diagnosis of CPM (Figure 1A, 1B). Although he remained conscious, complete paralysis of the facial, bulbar and limb muscles developed, with only the blinking and movement of the eyeball preserved. He was transferred to the general ward 3 weeks later and transferred to the Department of Rehabilitation Medicine 2 months later. His transfer was delayed due to fever and anal abscess. Follow-up brain MRI scan repeated one year later showed encephalomalacia at the site of the high-intensity T2 signal lesion in the pons (Figure 1C, 2D) (9)

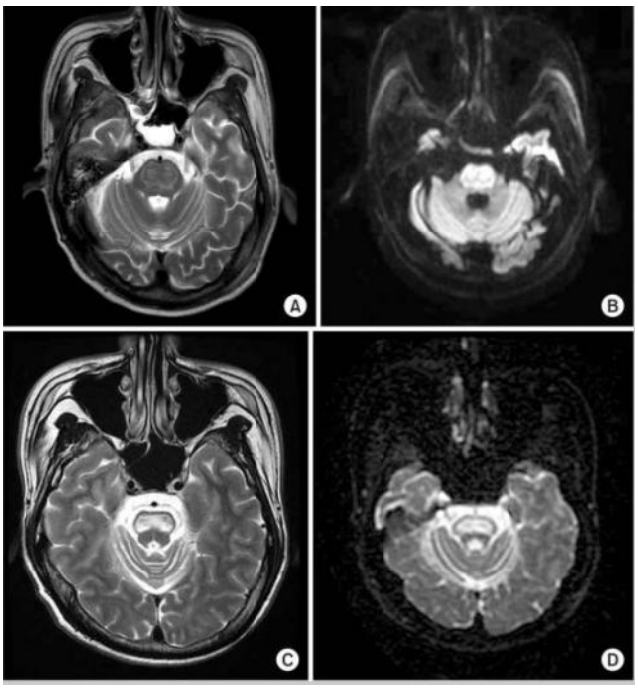

Figure 1. (A, B) Brain MRI shows T2- and diffusion-weighted imaging high signal intensity lesion in the bilateral pons, osmotic demyelination syndrome in pons 10 days after onset of symptoms. (C, D) Encephalomalacia at the site of initial high signal intensity lesion in pons after 1 year

Noor A & et al in their case report describe the case of a 47-year-old man who was transferred to the Medical College of Ohio Hospital due to sudden onset of clumsiness and difficulty walking.

Three weeks earlier, he had been admitted to a hospital after a period of binge drinking, fever, cough, and vomiting. Right upper and lower lobe pneumonia and acute gastritis were diagnosed. On admission, his serum sodium level was 123 mEq / L (reference ranges are shown in parentheses) (140-148 mEq / L). Hyponatremia was treated and corrected to 133 mEq / L within 24 hours and after 48 hours to 140 mEq / L.

The results of other laboratory studies at that time were as follows: white blood cell count, 17 × 109 / L (4-12 × 109 / L); serum urea nitrogen, 18 mg / dL (7-21 mg / dL); serum creatinine, 1.0 mg / dL (0.9-1.1 mg / dL); serum potassium, 3.4 mEq / L (3.6-5.2 mEq / L); serum chloride, 80 mEq / L (100-108 mEq / L); bicarbonate, 21 mEq / L (21-32 mEq / L); and vitamin B 12, 633 pg / mL (157-1059 pg / mL). Cerebrospinal fluid examination findings were normal. The chest radiograph showed evidence of pneumonia affecting the right upper and lower lobes. Sputum culture produced a combination of pneumococcus and Klebsiella pneumoniae. Blood cultures were negative. After intravenous administration of antibiotics, his pneumonia resolved, but pronounced appendicular and axial ataxia were noted. The patient was then transferred to Medical College Hospital for evaluation of these neurological findings. Upon examining it, he was fully conscious; he was time, place and person oriented; and he understood all the commands but had dysarthria. Force was normal in all limbs. He was clumsy when performing toe-nose and heel-knee maneuvers on both sides. His ability to perform rapid alternating movements was affected in both hands. A slightly reduced vibration sensation was noted at his ankles, but otherwise the sensations were normal. Deep tendon jerks were 2+ and symmetric, and Babinski's sign was negative bilaterally. The patient was unstable when sitting up and was markedly ataxic as he walked with a wide base gait. Magnetic resonance imaging (MRI) of the brain showed an area of increased signal in the pons on T2-weighted images and a decrease in signal intensity on T1-weighted images (**Figure 2 y 3**).

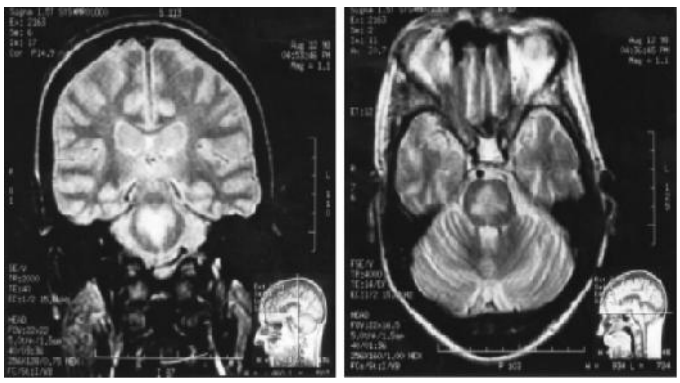

Figure 2. T2-weighted magnetic resonance images at the level of the pons. Coronal (left) and axial (right) views show an area of hyperintensity in the central pons. Note the bat's wing configuration in the coronal image (left) and the triangular shape in the axial image (right)

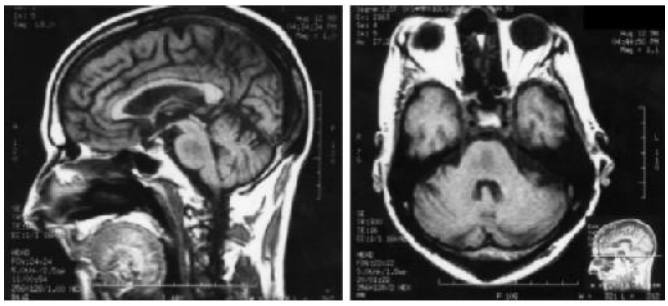

Figure 3. T1-weighted magnetic resonance images. Sagittal (left) and axial (right) views show hypointensity in the base of the pons. Note that the cerebellum and middle cerebellar peduncles are not involved (right)

The patient has a long-standing history of alcohol abuse and hyponatremia complicating an acute medical illness. Hyponatremia corrected and was followed by the appearance of new neurological signs; a pontine lesion on MRI was characteristic in appearance and location. These features support the diagnosis of CPM. This case was unusual in at least 2 respects. First, isolated cerebellar symptoms are a rare manifestation of CPM (10). Another case report by Robert C Tung describes a 56-year-old man with a history of alcohol abuse who was admitted to our trauma service after a motor vehicle collision, where he was a passenger. The patient was found to be unresponsive with a Glasgow coma score of 3 (E1, V1, M1). Initial brain images demonstrated a small subdural hematoma along the falx cerebri as well as the frontal convexities. Magnetic resonance imaging (MRI) of the cervical spine demonstrated severe posttraumatic changes at the C5-C6 level with no evidence of cord compression. The patient regained consciousness 36 hours after admission. His neurological examination a week later was remarkable for an awake patient capable of blinking on command.

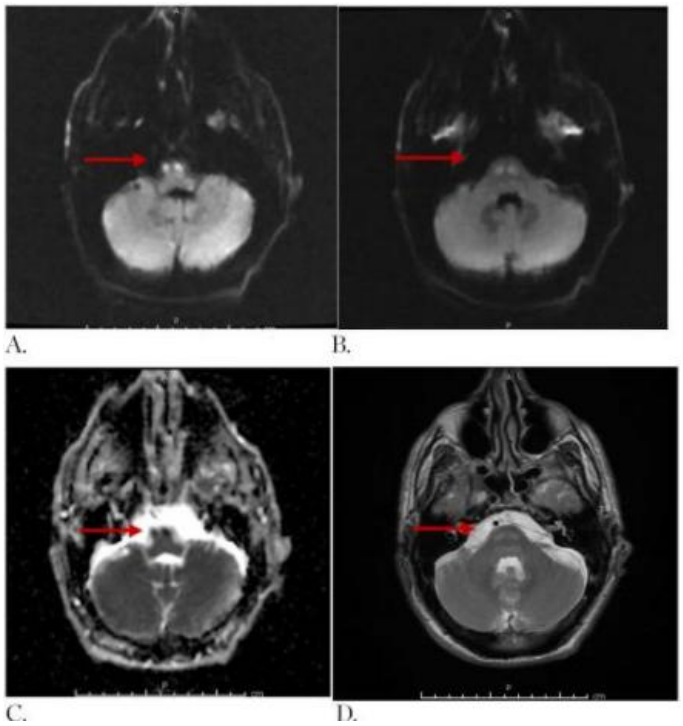

Figure 4. Axial MRI of the brain obtained 36 hours after admission demonstrates (by red arrows) symmetric areas of increased signal in diffusion weighted imaging (A, B) with decreased signal in apparent diffusion coefficient (C) and high T2 signal (D) at the level of the medullary pyramids and caudal pons involving predominantly white matter tracts

The pupils were equal, and reactive and extraocular movements had restricted bilateral abduction. The remaining bilateral vertical eye movements were intact. He had a bilateral facial, lingual, and palatal weakness that affected joint, swallowing, and respiratory capacity. He presented with flaccid tetraplegia with hyperreflexia and the presence of bilateral Hoffman signs, and required complete ventilatory support through tracheostomy. His neurological examination was consistent with "cloistered syndrome", suggestive of a lesion at the level of the pons. MRI of the brain demonstrated symmetric areas of subtle diffusion restricted in the pons and medulla involving corticospinal and corticobulbar fibers, suggesting a traumatic axonal lesion of the brainstem (Figure 4) (11).

DISCUSSION

During the 1950s and 1960s, the terms central pontine and extrapontine myelinolysis (joint osmotic demyelination syndrome) were used for the first time, which are demyelinating diseases characterized by histologically presenting as symmetric lesions in the white matter of the cerebellum., thalamus, globus pallus, putamen and, less commonly, in the peripheral cortex, hippocampi and lateral geniculate bodies; and it commonly occurs in malnourished patients, transplant recipients, or chronically debilitated patients, especially in those with electrolyte imbalances with rapid hyponatremia correction, as evidenced in the review of cases previously carried out. Being a demyelinating disease, it progressively affects the patient's motor capacity, completely affecting her quality of life, evolving to the development of a locked-in syndrome or pseudocoma, which ends in the majority of cases in the patient's death. Thus, it is necessary to implement a rapid, effective and accurate diagnosis by neurology and with the pertinent imaging tests. Therefore, the performance of magnetic resonance imaging in recent years has been a tool to confirm this diagnosis, taking into account the status of the results found through it. In his case report, Delgado presents a 47-year-old patient with osmotic demyelination syndrome secondary to a correction of hyponatremia who undergoes an MRI due to neurological deterioration and which findings were observed by linear images of subcortical location that follow the trajectory of the cerebral cortex, shown with diffusion restriction and hyperintense in T2 and FLAIR sequences, located in the left frontal lobe, both precentral gyrus, left insula and left parietal lobe, as well as in both thalamus. Therefore, taking into account the symptoms associated with the locked-in syndrome that includes quadriplegia, spasticity, hyperreflexia and pyramidal signs due to the injury of the corticospinal tracts at the level of the pontine base; anarthria and aphagia, due to injury to the corticobulbar fibers that travel in the dorsal portion of the bridge near the tegmento-basal junction, and horizontal gaze paralysis, due to injury to the pontine paramedian reticular formation, and the confirmatory images found In magnetic resonance imaging, which, due to its ability to demonstrate the brainstem without artifacts from the adjacent bone structures and its great sensitivity to detect changes in the white matter, constitutes the method of choice, it is possible to make a correct diagnosis. Fernandez et al. Describe in their case report the findings evidenced in a pregnant patient, who after a cesarean section without complications developed symptoms suggestive of pontine myelinolysis, for which they proceeded to perform magnetic resonance imaging in which it was evidenced At the level of

the pons and midbrain, bilateral hypointense symmetric lesions in T1 and hyperintense in FLAIR, likewise at the supratentorial level in the basal ganglia, similar lesions were observed in both sequences and with a bilateral and symmetric distribution. In the sagittal slices in the T2 sequence in projection of the corpus callosum, there was a hyperintense lesion (13) which confirmed the diagnosis of the clinical hand, which makes evident the importance of the rapid performance of this imaging test that can prevent the evolution of the pathology and therefore the complication of the neurological and motor status of the patient. The use of computed tomography (CT) as a method has been widely used in recent years for the diagnosis of this pathology, however, according to the series of cases reviewed, a higher degree of effectiveness has been evidenced in magnetic resonance imaging, because this evidence a marked hyperintensity in affected regions early, with a view to an eventual therapeutic intervention within the first 12 hours after the onset of symptoms (14), according to "The guidelines of the American Academy of Neurology ", As mentioned by Lara and others in their case presentation, that is, magnetic resonance imaging shows more relevant findings and therefore suggests prompt intervention by the patient and proper management of the same. Gamarra et al., In their case report, present a 61-year-old female patient who was admitted to a hospital with a clinical picture consisting of hyponatremia, which was corrected, and subsequently (7 days) developed progressive quadriparesis, dysarthria and finally dysphagia. Therefore, a computed tomography scan was requested, which did not show significant findings, however, 15 days later the patient presented an obvious neurological deterioration for which a magnetic resonance was performed, which showed hyperintensity at the level of the central portion of the pons., the basal ganglia and both thalamus in T2-weighted sequences and attenuated flow inversion recovery (FLAIR), and T1-weighted hypointensity, with restriction in the diffusion technique, which confirmed the diagnosis of osmotic demyelination, despite This, the patient developed cloistered syndrome and finally died, so it was concluded that the CT it is much less sensitive than MRI for the imaging of ODS, as the pons may be hypodense by artifact of the nearby petrous bone. The areas affected by myelinolysis are usually identified as hypodense lesions in the study without contrast, reflecting edema. (15)

Conclusion

The Cloistered Syndrome is a neurological disease caused by a lesion in the ventral portion of the pons, generally secondary to the development of pontine myelinolysis that manifests clinically with tetraplegia, anarthria, preservation of the level of consciousness and the ability to express through vertical eye movements or blinks, among other symptoms. Due to the fact that this develops in patients who have subsequently corrected hyponatremia, it is of great importance to correctly and quickly diagnose it, since patients who are not treated properly tend to develop more severe complications that lead to death. In recent years, computed tomography was known as the confirmatory imaging test, however, it has been shown that magnetic resonance imaging reveals more accurate, clear and suggestive findings of pontine myelinolysis, therefore, in the company of the clinic, it constitutes the diagnostic method more appropriate, which makes its rapid implementation necessary in patients with this pathology, in order to provide a correct approach to it, therefore, it is necessary that health personnel, mainly radiologists, are highly trained in identifying these Radiological characteristics for the diagnosis of pontine myelinolysis as a cause of cloistered syndrome.

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