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Case Report

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Primary Side Sclerosis, Case Report and Bibliographic Review

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Abstract

Introduction: *Primary lateral sclerosis is a rare disease that involves the upper motor neuron, producing a bulbospinal spasticity. The course of the disease is insidious and progressive, usually beginning with the lower extremities, and subsequently becoming a tetrapyramidal syndrome. Being a rare disease, the diagnosis in most cases is exclusion, having to study the patient extensively, in a clinical manner, including a thorough clinical history, laboratory and with relevant cabinet studies.*

Clinical Case: *This is a male patient who started his clinical picture about a year ago with weakness in the left pelvic limb, later accompanied by pain and paraesthesia, manifesting the same symptoms later in the contralateral leg and upper left limb. Currently, hypoesthesia of the index and middle toes of the right foot is added, moderate tremor in the left arm, with overlapping of the middle finger over the ring of said hand. He has an inability to lift light objects for short periods of time, as well as fatigue in short periods of time when performing daily activities, which greatly limits his daily life.*

Conclusions: *Motor neuron diseases are divided into two groups, and in the case studied, the upper motor neuron is exclusively affected. Being a rare disease, with a low incidence, multiple differential diagnoses will be considered before concluding this, considering a diagnosis of exclusion. The natural history of the disease will always have a bleak outcome, with a poor prognosis for life and function, despite the measures taken to modify its course.*

Keywords: Motor Neuron, Higher, Spastic, Resonance

Introduction

Primary lateral sclerosis is a rare disease that involves the upper motor neuron, which is characterized by a progressive bulboespinal spasticity, with a selective degeneration of pyramidal neurons located in the precentral gyrus [1]. Primary lateral sclerosis constitutes approximately 1% to 4% of all patients with motor neuron diseases [2]. The onset of the disease is insidious, with a slow and progressive spastic paralysis, usually starting at the lower extremities, to subsequently become a tetrapyramidal syndrome. On average, it is estimated that patients have a prognosis of life greater than 10 years from the onset of symptoms. Hyperreflexia, moderate weakness, dizziness, lack of coordination can also manifest within the clinical picture. On physical examination, spasticity, increase in osteotendinous reflexes, and as a predominant sign, stiffness can be found [2,3]. The diagnosis is usually made clinically, based on magnetic resonance imaging, among other studies, to rule out other pathological entities, evidencing in this I study a marked atrophy in the primary motor cortex [3]. In some studies, diffusion magnetic resonance imaging has been used, showing a functional increase in cerebral axonal activity, especially the cerebellar brain, which could explain an adaptive process through of functional neuroplasticity, however, the prognosis of the disease remains unfavorable and irreversible despite these changes [4]. Although there are some diagnostic criteria for the disease, being a rare entity, not all of the conditions are always met. Inclusions for it, so it is usually a diagnosis of exclusion [5].

Table 1: Diagnostic criteria proposed by Pringle and Cols

• Clinicians:

1.- Insidious onset of spastic paresis, usually starting in the lower extremities, but occasionally in the bulbar system or in the upper limbs.

2.- Start in adulthood, usually in the fifth decade of life or later.

3.- Absence of family history.

4.- Gradually progressive course.

5.- Duration greater than or equal to three years.

6.- Clinical findings generally limited to dysfunction of the corticospinal system.

7.- Symmetric distribution, with final development of severe spinobulbar spastic paresis.

• Laboratory (for differential diagnosis).

1.- Normal serum blood chemistry including vitamin B12 levels.

2.- Negative serology for syphilis and endemic areas for Lyme disease and HTLV-1 virus.

3.- Normal CSF parameters including absence of oligoclonal bands.

4.- In the majority of patients absence of potential denervation in

EMG, occasional fibrillation and increased insertion activity in few muscles. Absence of compressive lesions of the cervical cord or foramen in MR imaging

• Additionally suggestive of primary lateral sclerosis.

1.-Preserved bladder function.

2.- Absence or very long latency in cortical evoked motor responses in the presence of normal peripheral evoked stimuli combined with muscle action potentials.

3.-Focal atrophy of the precentral gyrus in MR.

4.- Disminución del consumo de glucosa en la región pericentral en PET.

Martínez, M. & Leyva, R. Esclerosis lateral primaria (reporte de un caso clínico y revisión de las manifestaciones clínicas). Revista mexicana de neurociencia. (2001). Vol. 2(3). Pp. 165-167. [6].

Clinical case

This is a 42-year-old male patient, who began his clinical picture about a year ago, when he was parading, I noticed some weakness in the left pelvic limb, progressing over the course of days to diffuse pain and local paresthesia, for later having the same problem in the contralateral leg and then a left upper limb in all its extension, progressively to date. Currently, in addition to the aforementioned, he presents hypoesthesia of the index and middle toes of the right foot, stiffness in all the arches of the movement of the affected limbs, moderate tremor in the left arm, with overlapping of the middle finger over the ring of said hand. Symptomatology is exacerbated in the morning. He has an inability to lift light objects for short periods of time, as well as fatigue in short periods of time when performing daily activities, which greatly limits his daily life. It denies heredofamilial antecedents of importance, as well as surgeries, drug addictions or known diseases.

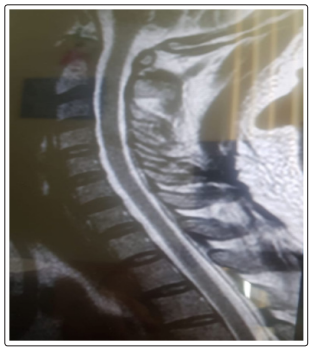

Image 1: Cervical spine magnetic resonance where there is a normal morphology and without compromise of the spinal cord

Discussion

Primary lateral sclerosis is a rare disease that affects the upper motor neuron of the primary motor area (precentral gyrus), characterized by having an insidious onset in people without known risk factors, with a clinical picture in which a spastic paresis of lower limbs of onset that can be generalized to produce a tetraparesis, which reflects a dysfunction in the corticospinal tracts, the severity of which increases progressively over the years and which is usually a disease of long evolution.

Motor neuron diseases are divided into those that affect the upper and lower, in this case we focus on the first case, which are within the cerebral cortex and send axons that form the pyramidal pathway, to later decide and finally connect to the spinal cord. The manifestations secondary to the upper motor neuron lesion are the following: spastic paralysis, amyotrophy (due to disuse), absence of fasciculations, exalted myotatic reflexes and extensor plantar response.

Being a disease with a low incidence and few reported cases, it should never be considered as a diagnosis of first instance, and other differential diagnoses must be ruled out, so over time various diagnostic criteria have been defined to specify the disease, however , as it was well mentioned, it is a rare disease, so a definitive consensus has not been created for the diagnosis and scrutiny of said disease, however, we can direct our diagnostic approach through any of these, such as criteria of pringle et al, through a well-established clinic, conducting a thorough medical history, laboratory studies, ranging from general analyzes such as blood count, blood chemistry, functional tests, to cytological examinations of cerebrospinal fluid, quantification of vitamin B12 in serum, treponemal tests, as well as cabinet studies such as l to electromyography and magnetic resonance imaging, where the absence of other pathological entities is verified and in this last study a marked cortical atrophy of the precentral gyrus. Despite the aforementioned, it should be borne in mind that the disease may not follow a specific pattern within its natural history, so the presentation and sequence of them can vary [6].

Conclusion

Motor neuron diseases are divided into two groups, and in the case studied, the upper motor neuron is affected exclusively, differentiating one from the other by the presentation and clinical manifestations. Being a rare disease, with a low incidence, multiple differential diagnoses will be considered before concluding this, considering a diagnosis of exclusion, and thus, a thorough medical history with adequate neurological examination should be performed, performing laboratory studies and cabinet relevant to that situation. Although it is a disease of long evolution and with a longer life expectancy than that of amyotrophic lateral sclerosis, the natural history of the disease will always have a bleak outcome, with a poor prognosis for life and function, taking into account account that there is no cure for the disease other than supportive treatment.

References

1. Tomik, B. Anna C, Szczudlik A (2008) Pure primary lateral sclerosis, case reports. Clinical Neurology and Neurosurgery 110: 387-391.
2. Jeffrey, M Richard, J Mazen, M Kay, M Mitzumoto H (2015) Primary lateral sclerosis. Neurologic clinics 33: 749-760.
3. Agosta, F Canu, E Innugi, A Chio, A Riva, et al. (2014) Resting state functional connectivity alterations in primary lateral sclerosis 35: 916- 925.
4. Meoded, A Morrissette, A Katipally, R Schanz, O Gotts, et al. (2015) Cerebro-cerebellar connectivity is increased in primary lateral sclerosis. Neuroimage: Clinical7: 288-296.
5. Santosh R, Rana S (2018) Demographics and clinical characteristics of primary lateral sclerosis: case series and review of literature. Neurodegenerative Disease Management 8: 17-23.
6. Martínez M, Leyva R (2001) Primary lateral sclerosis (report of a clinical case and review of clinical manifestations). Mexican journal of neuroscience 2: 165-167.