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Case Report

Therapeutic Plasmapheresis in Optical Neuromyelitis

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Abstract

A 35-year-old, male with gradual bilateral decrease in visual acuity of 5-weeks history of evolution. The campimetry showed 0% of visual field in the left eye and 84% in the right eye, with temporal hemianopsia, imaging studies were performed to rule out any lesion in optic chiasm. Both computerized tomography (CT) and magnetic resonance imaging (MRI) of the brain showed inflammation at the intersection of the optic nerve to the brain (optic neuritis). Likewise, an immunological study was performed to detect the acuaporin-4 antibodies (antiAQP4), being this result positive; so, the diagnosis of optical neuromyelitis (NMO) was concluded. The patient was treated first with steroids at high doses, but no improvement was observed. Therefore, based on the treatment guidelines of the American Apheresis Society (ASFA), Therapeutic Plasma Exchange (TPE)/therapeutic plasmapheresis was decided, giving positive results from the first session.

Keywords: Neuromyelitis Optica; Autoantibodies; Plasmapheresis

Introduction

Neuromyelitis optica or Devic's disease, is an autoimmune disorder, recurrent, characterized by demyelination of the Nervous System Central (CNS), which mainly affects the optic nerve and spinal cord, mediated by the production of antibodies against aquaporin-4 (anti-AQP4 or NMO-IgG), an autoantibodie which has a very important role in CNS homeostasis [1]. NMO was previously considered a form of multiple sclerosis (MS); however, there are significant clinical, laboratory, prognosis, and response to treatment differences between these two entities. Within the criteria for the diagnosis of NMO, there must be 2 major criteria such as optic neuritis and acute myelitis and 3 minor criteria: nuclear magnetic resonance imaging (MRI) that does not meet criteria for MS, spinal cord MRI with an extended lesion to 3 or more contiguous vertebral segments and presence of anti-AQP4 in serum, although it is required that the two major plus two minor criteria are met for definitive diagnosis, both optic neuritis and acute myelitis can occur simultaneously or not, so each case should be studied individually, since misdiagnosis predisposes to incorrect treatment, higher rate of relapse or recurrence and a worse prognosis [1,2].

A complete neurological exam is important to performed in cases of changes in visual acuity, it made include the evaluation on mental status, cranial nerve exam and coordination exam. Muscle strength and deep tendon reflexes in response of stimuli are also important part of the neurological examination. The visual campimetry or field is a test that measure the total area in wich the patient is able to see objects around affixed point. The optical coherence tomography (OCT) is a non-invasive imaging test that

uses light waves to take cross-section pictures of the retina layers to determine retinal or optic nerve damage. In many cases spinal tap is necessary in order to associate the type of pathology [6].

Case Report

A 35-year-old male, with no significant pathological history, consulted for presenting total loss of vision in the left eye and decreased visual acuity in the right eye, which occurred gradually 5 weeks of evolution. Haematological and serum chemistry examinations with normal values, positive anti-AQP4 antibodies and negative anti-MOG antibodies, chest CT scan and CT of abdomen with contrast medium, without alterations, upper gastrointestinal endoscopy revealed hiatal hernia. MRI of orbits reveals evidence of increased diameter of the optic nerve left eye; as well as an increase in loss of membrane continuity with a slight tortuosity in relation to the right eye (Image 1). CT and brain MRI report inflammation in the crosslinking of the optic nerve to the brain, with greater accentuation of the left eye (Image 2). Campimetry results reveal an unfavorable outcome, with a visual field index of 0% in the left eye and 84% in the right eye; neurological visual field suggesting temporary hemianopsia due to possible damage to the optic chiasm. The first-line treatment used was methylprednisolone 1 g for 5 days intravenously; at high doses, when there is no favorable response, RPT is performed, supported by the international guidelines of ASFA 2019 [3].

Image 1: MRI of Orbits: thickening of the optic nerve, left eye; and loss of membrane continuity and slight tortuosity (→).

Image 2: Brain MRI, axial plane: inflammation in crosslinking of the optic nerve to the brain, with greater accentuation of the left eye (→).

The first-line treatment used was methylprednisolone 1 g for 5 days intravenously; at high doses, as there is no favorable response, TPE is performed, supported by the international guidelines of ASFA 2019; [3] it was decided to perform 5 procedures on alternate days, with campimetry before and after RPT on a monthly basis, giving favorable results with recovery of visual acuity from the first session. The patient in his last session recovers 78% of visual acuity in the left eye and with an increase of 8% more, in the right eye (92%). Currently, he is on rituximab 20 mg every 6 months and after 12 months of finishing therapy with RPT the patient has not presented relapses, nor symptoms that suggest a picture of acute myelitis.

Discussion

NMO is an autoimmune, recurrent, demyelinating disease with CNS involvement, which was initially thought to be a variant of MS, but significant differences are now known between the two [1,2]. The largest number of cases have been reported in women [2]. NMO has clinical, imaging and laboratory criteria for diagnosis given by the Mayo Clinic and subsequently coined by Lennon., *et al*. who claim that positive anti-AQP4 serum autoantibodies have a sensitivity of 76% and a specificity of 94% for the diagnosis of NMO [1]. Major criteria may be presented simultaneously (monophasic) or at intervals of weeks, months, years to decades (outbreaks) [4]. In our case, the patient presented clinical optic

neuritis, as the first and only event, supported by brain MRI that shows inflammation in the crosslinking of the optic nerve to the brain, without typical MS lesions and positive anti-AQP4 antibody, without symptoms of acute myelitis, thus diagnosing a picture of NMO. These findings, unfortunately, do not rule out that the patient may not present a relapse or recurrence in the future, so it is very important a diagnosis, timely treatment of a possible relapse and above all, a maintenance therapy to avoid these possible future relapses. The first-line treatment is steroids at high doses such as methylprednisolone 1 g intravenously for 5 days, followed by descending regimens of oral prednisone, evaluating dose/response with the help of campimetry, in the case of our patient, he did not present any improvement, so he underwent the second or third line treatment that is the RPT [3-5], showing significant improvement from the first session. Patients who respond positively to TPR in future relapses should be treated in the same way, with TPR being the first-line treatment in these cases. To avoid future relapses, patients are treated with immunosuppressants [5], currently, our patient is treated with Rituximab, and after 12 months of his last RPT, he has not presented relapses. In conclusion, NMO is a disease with a poor prognosis, with which patients worsen with each recurrent episode of the disease, and may present total blindness due to optic neuritis or die from respiratory failure secondary to myelitis; However, the diagnosis and timely treatment of this pathology will allow the patient to have a better quality of life and have a lower rate of relapse [4].

Summary

An adult male with a gradual bilateral decrease in visual acuity over 5 weeks of evolution. Campimetry was performed, showing 0% of visual field in the left eye and 84% in the right eye with temporal hemianopsia, so imaging studies are performed to rule out lesion in optic chiasm. Both computed tomography (CT) and brain magnetic resonance imaging (MRI) show signs of optic neuritis. Likewise, an immunological study is performed for the detection of autoantibodie: anti-aquaporin 4 antibodies (anti-AQP4) being positive, so it is treated as neuromyelitis optica (NMO), was treated with steroids at high doses, but no improvement was observed, so supported by the treatment guidelines of the American Society of Apheresis (ASFA), a therapeutic plasmapheresis was performed, giving positive results from the first session.

Bibliography

1. Wingerchuk D M., *et al*. "Revised diagnostic criteria for neuromyelitis optica". *Neurology* 66.10(2006): 1485-1489.

2. Chiquete E., *et al*. "Neuromyelitis optica". *Revista Mexicana de Neurociencia* 11.3 (2010): 234-239.

3. Padmanabhan A., *et al*. "Guidelines on the Use of Therapeutic Apheresis in Clinical Practice – Evidence-Based Approach from the Writing Committee of the American Society for Apheresis /ASFA: The Eighth Special Issue". *Journal of Clinical Apheresis* 34.3 (2019): 267-268.

4. Bravo-Lizcano R., *et al*. "Devic's neuromyelitis optica". *RevClinMedFam* 9.2 (2016): 114-118.

5. Bruscolini A., *et al*. "Diagnosis and management of neuromyelitis optica spectrum disorders - An update". *Autoimmune Review* 17.3 (2018): 195200.

6. Oertel FC., *et al*. "Retinal Optical Coherence Tomography in Neuromyelitis Optica". *Neurology: Neuroimmunology and Neuroinflammation* 8.6 (2021): e1068.