Neuromyelitis optica spectrum disorder presenting with visual loss and apathy: A case report

Carolina Madeira[1], Gonçalo Godinho[1], Renato Santos-Silva[1], Ana Paula Pina, Manuel Falcão[1,2,3], Olinda Faria[1], Fernando Falcão Reis[1,2,3]

Background and Purpose

Neuromyelitis optica (also known as Devic's disease or Devic's syndrome) is an uncommon disorder in pediatric age group, and is characterized by acute or subacute optic neuritis and transverse myelitis.

We report a case of neuromyelitis optica spectrum disorder in a 17 years old teenager.

Clinical Presentation

A seventeen-year old female presented with bilateral progressive diminished vision, in the last four days. During anamnesis and examination, she was apathetic and collaborated poorly. She reported occipital headache, upper and lower left limb numbness and nausea in the past week. Her medical history was unremarkable.

Neurological examination was normal.

Clinical Testing

Initial exam	OD	OS
Visual acuity	LP	LP
Pupils	No APD	
EOM's	Normal	Normal
PIO (mmHg)	18	18
Ocular Fundus	Normal	Normal

Legend: OD - right eye; OE - left eye; LP - light perception; APD - afferent pupillary defect; EOM - extraocular movements; PIO - intraocular pressure

- **Electrophysiological testing**

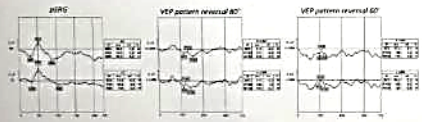

Figure 1 - The pattern visual evoked potential showed a decrease of 25% of P100 wave of the OD and a decrease in residual activity in the OS, with implicit time increased by 15ms in the OD. The pattern electroretinogram (pERG) showed a bilateral decrease of N95. These results were suggestive of bilateral optic neuritis.

- **Brain and Medular Magnetic Resonance Imaging (MRI)**

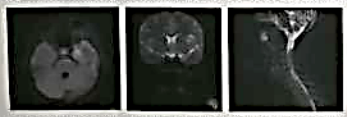

Figure 2 - The brain MRI revealed millimeter areas with an increased signal in T2 images after gadolinium contrast at cortical, sub-cortical, bilateral regions and of the optic chiasm. The medular MRI also showed multiple areas with a contrast enhanced consistent T2 signal that included the medullary cone, the largest encompassing 3 segments.

- **Lumbar puncture and laboratory work-out**
  - Extensive immunological study, including immunophenotyping was only positive for anti-MOG antibodies.
  - Blood and CSF serological and bacteriological tests were negative.

Plan and Treatment

The patient was started on methylprednisolone 1g/day for 5 days and then initiated oral prednisolone, which was gradually trapped. Rituximab was also initiated.

Evolution

Two months after the inaugural presentation, the patient had a great improvement, with resolution of neurological symptoms. At examination she presented a BCVA of 20/30 and 20/20 in OD and OS respectively.

- **Colour fundus photography**

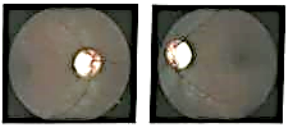

Figure 3 - Bilateral temporal optic disc pallor

- **Electrophysiological testing**

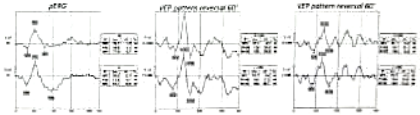

Figure 4 - The VEP showed a P100 wave with normal amplitude and a discrete implicit time increase (125 ms) in the OD and a normal P100 in the OS.

- **Visual Field and Optic Nerve OCT**

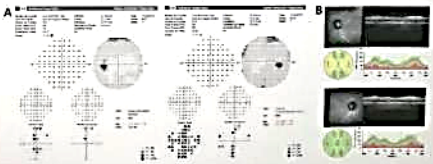

Figure 5 - A) Visual field test revealed in the OD unspecific points with sensitivity loss, and in the OS visual field was normal. B) Optic nerve OCT showed a retinal nerve fiber layer of thickness of 83 and 87 in the OD and OS, respectively. The OD presented a borderline thickness, with the temporal and nasal quadrants.

Conclusion

Our patient fulfilled the diagnosis criteria for NMOSD. She presented 2 core clinical characteristic (optic neuritis and acute myelitis) and additional MRI imaging characteristics. Diagnosis of this pathology remains a challenge, however distinguishing NMOSD from other demyelinating diseases is essential for prognosis and therapeutic management.

References

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2. Wingerchuk DM, Banwell B, Bennett JL, Cabre P, Carroll W, Chitnis T, et al. International consensus diagnostic criteria for neuromyelitis optica spectrum disorders. Neurology. 2015 Feb;85(2):177–89.