57ο Πανελλήνιο Οφθαλμολογικό Συνέδριο
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Θεσσαλονίκη Makedonia Palace Hotel

# Uncommon manifestations of Neuromyelitis optica (NMO) and challenges in the differential diagnosis: a case report

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ΠΑΝΕΠΙΣΤΗΜΙΑΚΟ ΓΕΝΙΚΟ ΝΟΣΟΚΟΜΕΙΟ ΗΡΑΚΛΕΙΟΥ

## Introduction

Neuromyelitis optica (NMO) is an inflammatory disease of central nervous system classically characterized by acute, severe episodes of optic neuritis and longitudinally extensive transverse myelitis, usually with a relapsing course. In this case report, we present an uncommon case of retinal ischemia and disc edema in a 30-year-old female with concomitant hearing loss and positive aquaporin-4 test.

## Materials and methods

The patient was evaluated on a daily basis. Visual acuity was determined using the standard Snellen optotype. Visual fields (VF) were recorded using the Humphrey automated perimeter in SITA standard 30:2. Serial widefield Retinal photographs were captured using the Optos widefield fundus camera. Retinal integrity was evaluated via SD OCT. A complete infectious disease, immunologic, coagulation factor and demyelinating disease workup were obtained upon admission.

## Case presentation

- A 30-year-old female presented at the emergency department reporting gradual-onset blurred vision in her right eye (OD), following an episode of hearing loss.
- In OD: Visual acuity was 1/10 with positive relative afferent pupillary defect, red desaturation and intraocular pressure of 19 mmHg. OS findings were unremarkable.
- Slit lamp examination of the anterior segment was normal, while fundus examination revealed stage IV optic disc edema with macular extension, retinal vessel tortuosity and dot-blot hemorrhages (Figure 1).
- Neurologic examination and brain MRI were reported unremarkable, whereas audiometry revealed bilateral hearing impairment.
- Visual field test showed a diffuse VF constriction OD and fluorescein angiography (FA) a delayed venous phase OD (Figure 2).
- The patient was admitted to the Neurology clinic, where she received intravenous corticosteroid therapy.


Fig 1: Colour widefield retinal photograph OD showing stage IV optic disc edema with macular extension, retinal vessel tortuosity and dot-blot hemorrhages

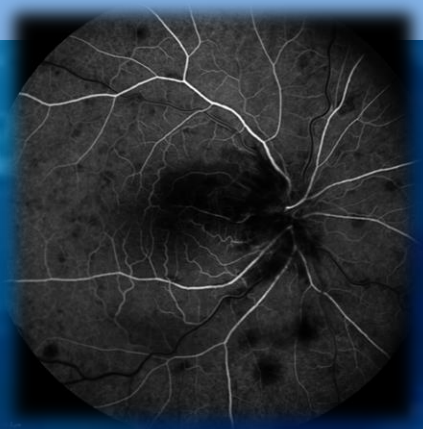

Fig 2: Fundus fluorescein angiography (FA) showing a delayed venous phase OD

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**Follow-up**

- One week post admission, no improvement was evident in her right eye with a visual acuity of 1/10, stable disc edema with diffuse retinal hemorrhages and inner retinal edema (Figure 3).
- Anti-AQP-4 Antibody titres came up positive.
- One month later, under oral methylprednisolone, visual acuity improved to 2/10 and fundus examination was indicative of both optic disc and inner retinal atrophy (Figure 4).
- Findings were indicative of neuromyelitis optica.

- However, as a second workup at that time for aquaporin-4 antibodies was negative a diagnostic challenge was posed.

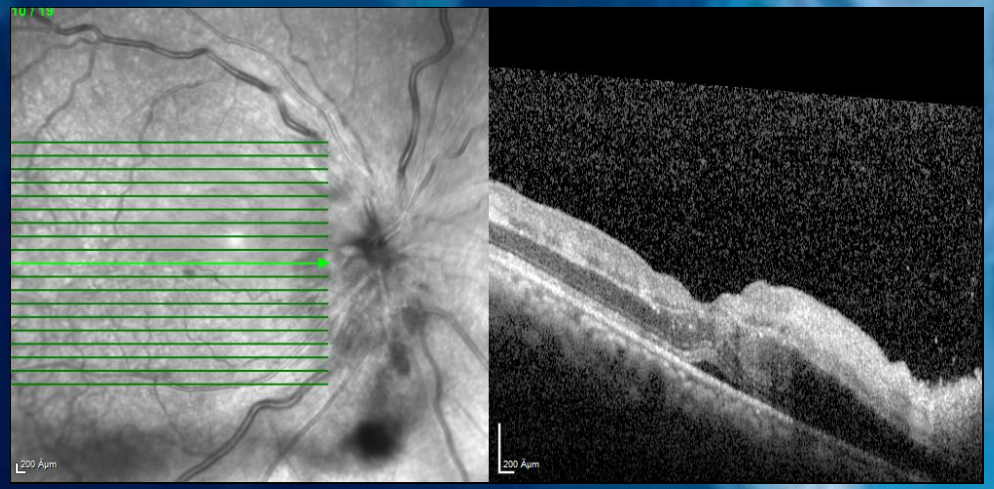

Fig 3: SD OCT scan OD showing diffuse inner retinal edema (one week post admission)

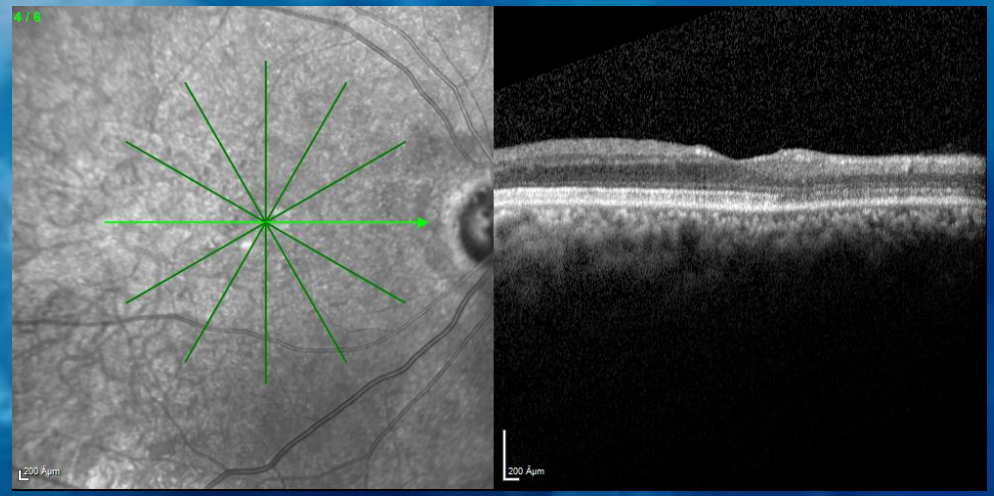

Fig 4: SD OCT scan OD showing diffuse inner retinal atrophy (one month follow-up)

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## Discussion

- Rare cases of retinal vascular occlusions associated with neuromyelitis optica disease spectrum (NMODS) have been reported in the literature (Lee et al., 2008).
- OCT findings elucidate NMODS associated inflammatory central retinal artery occlusion (Manogaran et al., 2016).
- Optic neuritis can be associated with retinal hemorrhages and ischemia (Chan et al., 2011); Kaushik et al., 2023; Kelly et al., 2019).
- Negative workup for aquaporin-4 antibodies may be associated with serum sampled soon after corticosteroid therapy (Takahashi et al., 2007).

## Conclusion

- Diagnosis of Neuromyelitis optica can be challenging in cases with atypical presentations.
- The prompt identification of NMO patients with such presentations may benefit these patients with institution of early treatment to reduce disability and prevent further attacks.

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