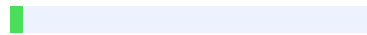




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Title

When All Tests Are Normal: Steroid Responsiveness as a Key Diagnostic Argument for Multiple Sclerosis in Pediatric Optic Neuritis

Abstract

Background:

Pediatric optic neuritis (ON) represents a diagnostic challenge, particularly when extensive investigations fail to identify a clear etiology. While imaging and laboratory findings are central to diagnosis, clinical reasoning and therapeutic response may play a decisive role.

Case presentation:

We report a 14-year-old girl presenting with acute unilateral visual loss of the right eye associated with headache and pain on eye movement. Clinical examination revealed anterior optic neuropathy with stage II optic disc edema.

A comprehensive ophthalmologic, neurological, infectious, and immunological workup was performed. Brain and orbital MRI showed inflammatory involvement of the right optic nerve without evidence of disseminated lesions. Cerebrospinal fluid analysis and extensive autoimmune and infectious testing were unremarkable.

Management and outcome:

High-dose intravenous methylprednisolone resulted in rapid and marked visual recovery, with resolution of optic disc edema.

Conclusion:

This case highlights that, in the absence of definitive paraclinical evidence, the **diagnosis of demyelinating optic neuritis** suggestive of multiple sclerosis may rely on clinical presentation and therapeutic response. Steroid responsiveness should be

considered a major diagnostic argument in such cases.

Keywords:

Optic neuritis; Pediatric; Multiple sclerosis; Steroid response; Diagnostic challenge

Introduction

2 Optic neuritis (ON) is an inflammatory disorder of the optic nerve frequently associated with demyelinating diseases, particularly multiple sclerosis (MS). In adults, diagnosis is often supported by magnetic resonance imaging (MRI) and cerebrospinal fluid (CSF) abnormalities demonstrating 4 dissemination in space and time.

However, pediatric ON differs in presentation and often lacks definitive paraclinical markers at initial evaluation. In such cases, the diagnostic process becomes more complex, requiring a careful integration of clinical, radiological, biological, and therapeutic data. Importantly, a rapid and significant response to corticosteroid therapy is recognized as a characteristic feature of inflammatory demyelinating optic neuritis and may contribute to the diagnostic orientation toward MS.

We report a case of pediatric optic neuritis in which all investigations were inconclusive, and the diagnosis relied primarily on clinical findings and a striking response to corticosteroids.

Case Report

A 14-year-old girl presented with acute visual loss in the right eye evolving over five days. The condition was associated with headache and pain on ocular movement.

Medical history

- Iron deficiency anemia for 18 months (poor compliance with treatment)
- Menstrual cycle disorders (hypermenorrhea and irregular cycles)
- Refractive amblyopia in the left eye
- Recent upper respiratory tract infection 10 days prior

Clinical Examination

- Visual acuity:
 - o Right eye: counting fingers at 2 meters (non-improvable)
 - o Left eye: 5/10
- Pupillary reflex:
 - o **1** Relative afferent pupillary defect in the right eye
- Intraocular pressure: normal bilaterally
- Anterior segment: normal
- Fundus examination (right eye):
 - o Stage II optic disc edema
 - o Blurred margins
 - o Papillary hyperemia
 - o Venous dilation and tortuosity
 - o Absence of hemorrhages or exudates
- Left eye: normal

Ophthalmologic Investigations

Fluorescein Angiography

- Papillary leakage
- Peripapillary diffusion
- Venous dilation

→ confirming inflammatory optic neuropathy

Optical Coherence Tomography (OCT)

- Increased optic disc thickness
- Reduction of physiological cupping
- Peripapillary RNFL thickening
- Macular OCT: normal

Visual Field

- Right eye: non-interpretable due to low visual acuity
- Left eye: learning test, arciform superior defect (to be confirmed)

Color Vision (Lanthony desaturated test)

- Blue-yellow (tritan) deficit
- Red-green (deutan) deficit

→ consistent with optic nerve dysfunction

Visual Evoked Potentials (VEP)

- Right eye: abolition of P100 response
- Left eye: delayed latency and reduced amplitude

→ suggesting:

- Severe pre-chiasmatic involvement (right eye)
- Subclinical involvement (left eye)
- Possible demyelinating process

Neuro-imaging

MRI (brain and orbits)

- T2 and FLAIR hyperintensity of the right optic nerve
- Gadolinium enhancement
- Involvement of intraorbital, retrobulbar, intracanalicular, and prechiasmatic segments
- No demyelinating lesions elsewhere
- No compressive lesion

Laboratory and Systemic Workup

Cerebrospinal Fluid

- Normal cytology
- Normal protein and glucose
- Negative culture

Biological Workup

- Mild anemia (Hb 10.7 g/dL)
- Elevated ESR (52 mm/h)
- Normal CRP
- Low ferritin

Infectious Workup

- HIV, HSV, CMV, EBV: negative or immunized
- Syphilis, Lyme, toxoplasmosis: negative
- Tuberculosis (Quantiferon): negative

Autoimmune Workup

- ANA, anti-dsDNA, ANCA: negative
- Anti-MOG: negative
- Anti-AQP4: negative

Differential Diagnosis

- Demyelinating optic neuritis (MS, NMOSD)
- Infectious optic neuropathy
- Autoimmune systemic disease
- Compressive optic neuropathy
- Hereditary optic neuropathy

All alternative diagnoses were excluded.

Management

- Oral antibiotics for associated sinusitis
- Intravenous methylprednisolone (1 g/day for 3 days)
- Oral corticosteroid relay

Outcome

Clinical evolution

- Visual acuity improved from CF → 9/10
- Pupillary reflex normalized
- Resolution of optic disc edema

Visual field

- Improvement with residual central scotoma

Discussion

Pediatric optic neuritis is often diagnostically challenging due to the absence of typical radiological and biological markers at onset.

In this case:

- MRI did not demonstrate dissemination in space
- CSF was normal
- Autoimmune and infectious workups were negative

However, several clinical arguments strongly supported a demyelinating etiology:

- Acute unilateral visual loss
- Pain on eye movement
- Optic disc edema
- 1 Relative afferent pupillary defect**
- Bilateral electrophysiological involvement

Most importantly, the rapid and marked response to corticosteroid therapy represents a well-recognized characteristic of inflammatory demyelinating optic neuritis and is considered an important diagnostic argument in favor of multiple sclerosis.

This case highlights a crucial clinical principle:

Diagnosis should not rely solely on paraclinical findings but must integrate clinical reasoning and therapeutic response.

Conclusion

This case illustrates the limitations of paraclinical investigations in pediatric optic neuritis.

Even **1** in the absence of definitive diagnostic findings, a demyelinating etiology suggestive of multiple sclerosis should be considered when clinical features are typical and when there is a strong response to corticosteroid therapy.

Close follow-up is essential to confirm the diagnosis over time.

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