



REVIEW

Optic neuritis in pediatric population: A review in current tendencies of diagnosis and management



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Abstract Optic neuritis is an inflammation of the optic nerve and may be related to different systemic conditions. The clinical presentation of this pathology usually includes sudden loss of visual acuity (VA) which may be unilateral or bilateral, visual field restriction, pain with eye movements, dyschromatopsia, a relative afferent pupillary defect and optic disk swelling. Optic neuritis in children has specific clinical features and a better prognosis than in adulthood. Although usually appears an underlying viral disease, the main concern for practitioners is the relationship of optic neuritis with multiple sclerosis. In addition to the classical techniques as magnetic resonance imaging (MRI), current tendencies of diagnosis for eye practitioners include new imaging devices as optical coherence tomography (OCT), useful to show a thinning of the retinal fibers layer (RFL) after the inflammatory episode. Regarding the management of these patients, short-term intravenous steroid dosages seem to be the best option to treat acute attacks characterized by a very poor bilateral VA.

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PALABRAS CLAVE

Neuritis óptica;
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Esclerosis múltiple;
Tomografía de
coherencia óptica

Neuritis óptica en una población pediátrica: revisión de las tendencias actuales de diagnóstico y tratamiento

Resumen La neuritis óptica es una inflamación del nervio óptico, que puede relacionarse con diferentes condiciones sistémicas. La presentación clínica de esta patología incluye normalmente pérdida súbita de agudeza visual (AV), que puede ser unilateral o bilateral, restricción del campo visual, dolor al mover los ojos, discromatopsia -defecto pupilar aferente relativo- y edema del disco óptico del disco óptico. La neuritis óptica en niños tiene características clínicas especiales, y un mejor pronóstico que en los adultos. Aunque normalmente parece

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una enfermedad vírica subyacente, la principal dificultad para los especialistas es la relación de la neuritis óptica con la esclerosis múltiple. Además de las pruebas clásicas como la resonancia magnética (RM), las tendencias actuales de diagnóstico para los profesionales de la salud visual incluyen nuevos dispositivos de imagen tales como la tomografía de coherencia óptica (TCO), que es útil para reflejar el adelgazamiento de la capa de fibras de la retina tras el episodio inflamatorio. Respecto al tratamiento de estos pacientes, la administración de esteroides intravenosos a corto plazo parece ser la mejor opción para tratar los ataques agudos, caracterizados por una AV bilateral muy reducida.

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Introduction

Optic neuritis is a primary inflammation of the optic nerve. Clinically, it can appear as an isolated condition or associated to a variety of systemic autoimmune disorders. Its incidence is of 1–5 per 100,000 per year.^{1–4} It mainly affects the Caucasian population, most often during the spring period, and especially to those living at high geographic latitudes.¹ Its origin is inflammatory and the pathophysiology of this condition includes a peripheral activation of T-cells that may cross the blood brain barrier causing a delayed type IV hypersensitivity reaction. This process leads to destruction of myelin,⁵ with also the involvement of the axon, something that can be confirmed by mean of an optical coherence tomography (OCT) examination. All this process has a negative impact on visual acuity (VA).⁶

The typical presentation of optic neuritis is a sudden and unilateral visual loss, with VA ranging from levels of 0.8 in decimal scale to no light perception (NPL). Ninety percent of patients refer pain of variable intensity, usually periocular, which is associated with ocular movements or may be a prelude of the episode of optic neuritis. The evolution of this conditions ranges from days to weeks and is common the association to phosphenes and flashes of light. Other clinical finding that can be also associated with optic neuritis is the Uhthoff' phenomenon that consists on a decreased VA in presence of a high body temperature or during physical exercise.¹ Likewise, other clinical features of optic neuritis are abnormal color vision, reduced contrast sensitivity function, visual field loss characterized by central, paracentral or altitudinal scotomas, and the presence of a relative afferent pupillary defect (RAPD), which is more evident in unilateral cases. In some cases, a mild to moderate Tyndall effect may appear in the anterior chamber or in the vitreous as well as a peripheral retinal periphlebitis (12% of patients). This last sign has been proposed as an indicator of a greater probability of developing multiple sclerosis (MS).⁷ In 36–58% of patients, optic neuritis is accompanied by papillary edema or optic head nerve swelling, whereas in the rest of cases inflammatory involvement occurs in the retrobulbar optic nerve portion and, therefore, the appearance of the optic nerve is normal.⁸

VA loss and pain are limited in time and occurs during the episode of acute inflammation. From this moment, the remyelination and proliferation of sodium channels in the neuronal segments begins, which is a process that can last even more than two years.⁹ The same duration has been reported for the reorganization of the cortical activation.¹⁰ Final VA is in relation with the severity of the initial VA

loss. Ninety-four percent of patients whose VA falls to less than 0.6 during the episode of optic neuritis recover their previous VA. Similarly, 64% of patients whose VA drops to perception of light (PL) recovered a VA of 0.5 or better.¹¹ However, VA tends to fluctuate after an episode of optic neuritis. Furthermore, it is frequent the perception of the Uhthoff' phenomenon, mentioned above, and the Pullfrich' phenomenon, which is exemplified with a pendulum seeming to describe an ellipse despite moving in a single plane, as a result of an asymmetric velocity of conduction between both optic nerves.¹² As RAPD is dependent on ratio of functional axons between both eyes, all these phenomena described may remain in most cases after the episode of optic neuritis. Likewise, pallor may remain normally in the temporal portion of optic nerve (Fig. 1).

The diagnosis of optic neuritis is complex and may lead to frequent errors.¹ Optic neuritis of the adult is associated with autoimmune (Lupus erythematosus, sarcoidosis, Behçet's disease) and infectious diseases (viral etiology, syphilis, tuberculosis or Lyme disease). The differential diagnosis of optic neuritis also includes optic neuropathies of different etiologies such us compressive, ischemic, hereditary, toxic and nutritional.



Figure 1 Optic disk temporal pallor after an episode of bilateral optic neuritis in a 13 years-old patient with a VA of 0.2 (decimal scale).

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