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Short communication

Neurotrophic keratopathy in a patient with familial amyloidosis☆

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ABSTRACT

Introduction: Familial amyloidotic polyneuropathy is a disease related to amyloid material deposits in the extracellular matrix that can affect many tissues, including the eye.
Case report: This case includes clinical data, a full corneal study report, and histopathological findings, as well as the treatment and follow up of bilateral neurotrophic keratopathy in a 33 year-old patient with familial amyloidotic polyneuropathy.
Discussion: Although amyloidotic deposits were also found in the conjunctival tissue, this is not a typical form of ophthalmic amyloidosis. The corneal wound in this case is a result of a corneal anesthesia due to the systemic polyneuropathy.

Queratopatía neurotrófica corneal bilateral en paciente con amiloidosis familiar bilateral

Palabras clave:
Amiloidosis familiar
Polineuropatía
Queratopatía neurotrófica

RESUMEN

Introducción: La amiloidosis familiar es una enfermedad caracterizada por el depósito extracelular de un material denominado amiloide que puede afectar a gran variedad de órganos, incluyendo el ojo.
Caso clínico: Se presentan datos clínicos, pruebas complementarias e histológicas, así como el tratamiento y seguimiento de un paciente de 33 años con úlcera neurotrófica corneal bilateral, debida a una polineuropatía amiloidótica familiar.
Discusión: Este caso no es la forma típica de presentación de la amiloidosis oftálmica, ya que la úlcera corneal es el resultado de la anestesia causada por la polineuropatía sistémica, aunque se encontraron depósitos de amiloide en el tejido conjuntival.

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Introduction

Amyloidosis is a disease of unknown etiology,[1] characterized by the extracellular sedimentation of a fibrillary substance of proteic origin in various tissues and organs, giving rise to functional and structural alterations therein.[1–3]

Familial amyloidotic polyneuropathy is related to deposits of the protein that fixes retinol, transthyretin, produced in the liver. This accumulates mainly in the peripheral nerves, producing polyneuropathies or myocardiopathies. It is a dominant autosomal disease, the most common mutation of which is the substitution of valine by methionine in position 30.[4] At present, the treatment yielding the best results is liver transplant[5]; however, severe ocular compromise is a long-term risk, with frequent vitreous opacities and glaucoma.[6] The literature contains few reports on neurotrophic keratopathy secondary to amyloidosis.

Clinic case report

Male, 33, who visited the Emergency Dept. with conjunctival hyperemia and itching in both eyes (BE) with one month evolution, without associating ocular pain. The patient had been treated with tobramycin cream during 15 days without clinic improvement. Pathological antecedents included celiac disease, psoriasis and familial amyloidosis requiring liver transplant and pacemaker implantation. Ophthalmological examination revealed visual acuity (VA) of 20/100 in the right eye (RE) and 20/50 in the left eye (LE). Biomicroscopy produced in both eyes marked ciliary hyperemia, with 1 mm diameter inferior paracentral corneal ulcer, with corneal thinning up to the deep stroma, predominantly in LE. Ulcer edges were well defined, without signs of acute infection (Fig. 1). No cells were evidenced in the anterior chamber, or hypopyon. Intraocular pressure was 14 mmHg in BE. Corneal sensitivity was assessed with a cotton swab, observing total anesthesia in BE. Optical coherence tomography (Cirrus HD-OCT 4.0, Carl Zeiss Meditec, Dublin, CA) demonstrated significant thinning in the ulcerated area, with a residual thickness of 360 μm in RE and 290 μm in LE. Treatment was initiated with cycloplegic eyedrops every 8 h, ofloxacin eyedrops every 2 h, erythromycin cream during the night and artificial tears every hour in BE. Due to the significant corneal thinning, 4 days later a lateral tarsoconjunctival bridge was made in the LE. At day 15, the patient exhibited partial cicatrization of the neurotrophic ulcers with diminished corneal thinning. VA was 20/160 in BE.

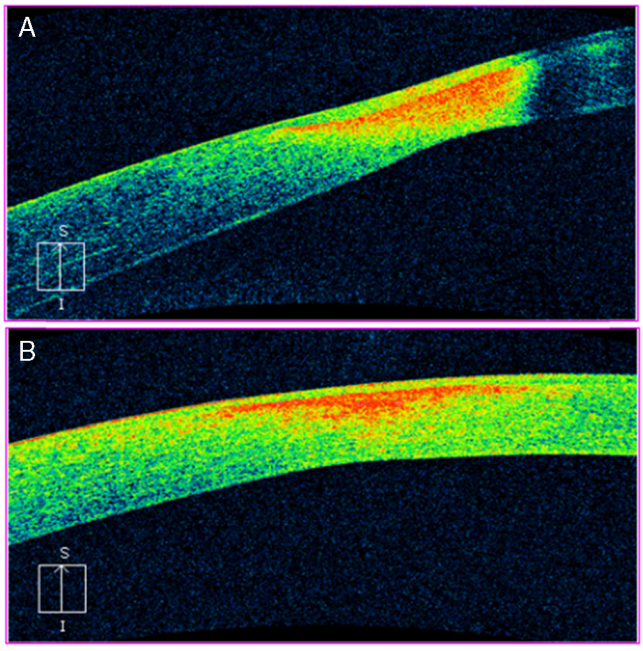

Fig. 2 – (A) Left eye neurotrophic ulcer OCT, 2 months post-diagnostic. (B) Left eye neurotrophic ulcer OCT 3 months post-diagnostic.

During the follow-up, OCT images were taken. At month 3, corneal thickness was 400 μm in LE (Fig. 2). At month 9, the tarsoconjunctival bridge was submitted to excision, when the ulcer was in the residual corneal leukoma phase (Fig. 3). Anatomopathological analysis of the bridge conjunctiva evidenced the presence of amyloid substance, both in hematoxylin eosin (Fig. 4) as well as apple green bi-refringence, characteristic of the substance after Congo red staining (Fig. 5), which confirmed the diagnostic.

Discussion

The present case is an infrequent occurrence of neurotrophic keratopathy due to corneal innervation compromise caused by familial amyloidotic polyneuropathy in which conjunctival compromise was additionally confirmed through anatomopathological study. Corneal-conjunctival amyloid deposits constitute an infrequent finding in the literature.[7–9]

Neurotrophic keratopathy is the result of trigeminal nerve compromise, which accounts for corneal innervation. The

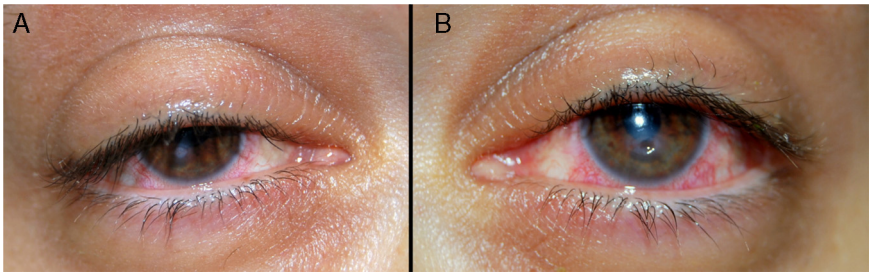

Fig. 1 – Pre-treatment neurotrophic corneal ulcers. (A) Right eye. (B) Left eye, showing greater depth of lesion.