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Short communication

Neurotrophic keratopathy secondary to trigeminal nerve aplasia in patient with Goldenhar syndrome☆

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ABSTRACT

Case report: A 4-year-old male diagnosed with Goldenhar syndrome, with an unremarkable ophthalmic history, develops a neurotrophic ulcer secondary to trigeminal nerve aplasia. It was treated with multilaminar amniotic membrane transplantation.
Discussion: Trigeminal nerve aplasia is not usually reported in Goldenhar syndrome. Therefore, it seems necessary to perform routine eye examinations, from an early age, to prevent serious complications associated with corneal anesthesia.

Úlcera neurotrófica secundaria a aplasia de nervio trigémino en paciente con síndrome de Goldenhar

Palabras clave:
Síndrome de Goldenhar
Aplasia nervio trigémino
Úlcera neurotrófica
Trasplante membrana amniótica
Hipoestesia corneal

RESUMEN

Caso clínico: Varón de 4 años de edad diagnosticado de síndrome de Goldenhar, sin antecedentes oftalmológicos relevantes, desarrolla una úlcera neurotrófica secundaria a aplasia de nervio trigémino que es tratada con trasplante de membrana amniótica multilaminar.
Discusión: En el síndrome de Goldenhar no suele estar descrita la aplasia de nervio trigémino como manifestación oftalmológica típica. Por tanto, parece necesario realizar controles

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oftalmológicos rutinarios y desde una edad temprana, para evitar la aparición de complicaciones graves asociadas a la anestesia corneal.

Introduction

The Goldenhar syndrome or oculo-auricular-vertebral dysplasia is a rare congenital malformation disease that prevents the adequate development of the structures derived from the first and second brachial arch during blastogenesis, with higher unilateral involvement and with a preference for males.[1–3]

An infrequently associated expression of this syndrome is trigeminal nerve aplasia, that could lead to the formation of neurotrophic ulcers which are generally severe in these patients.

Clinic case report

Male patient, 4, diagnosed at birth with Goldenhar syndrome due to exhibiting unilateral malformations on the left side consisting in hypoplasia in the pinna, mandibular, the first costal arches and collarbone, in addition to cleft palate, laringotracheobronchomalacia and left kidney agenesis. The patient did not exhibit relevant ophthalmological history.

The patient was taken to the Emergency Dept. due to reddening in the left eye (LE). The exploration of said eye revealed intense ciliary injection and a central and temporal corneal ulcer with a diameter of approximately 5 mm, with thickened edges and without infiltration. Total absence of sensitivity was verified (anesthesia) in the affected cornea, suspecting trigeminal nerve hypoplasia or aplasia. Topical treatment was initiated with antibiotic, corticoid and autologous serum.

A magnetic resonance (MR) study was decided to confirm the suspected diagnostic and describe possible associated anomalies. The imaging test showed cerebellar dysplasia, tentorial lipoma, hypoplasia of the brainstem and the left facial and cochlear-vestibular nerves in addition to confirming the ipsilateral trigeminal nerve aplasia (Figs. 1 and 2).

Two weeks later the ulcer continued torpid evolution and began to develop an abnormal hypertrophic cicatrization area (Fig. 3). It was decided to carry out a surgical examination as well as covering the epithelial defects with amniotic membrane.

During surgery the scar area debrided, evidencing a significant corneal thinning. Multilaminar amniotic membrane was transplanted (Fig. 4).

Twelve days after the above surgery the amniotic membrane detached after achieving full epithelialization of the ulcer. During follow-up, the persistence of a dense residual corneal leukoma was observed (Fig. 5) with a visual acuity of 20/50 in that eye.

In patients such as the present case, keratoplasty has a gloomy prognosis due to the possibility of severe post-surgery persistent epithelial defects, without mentioning the

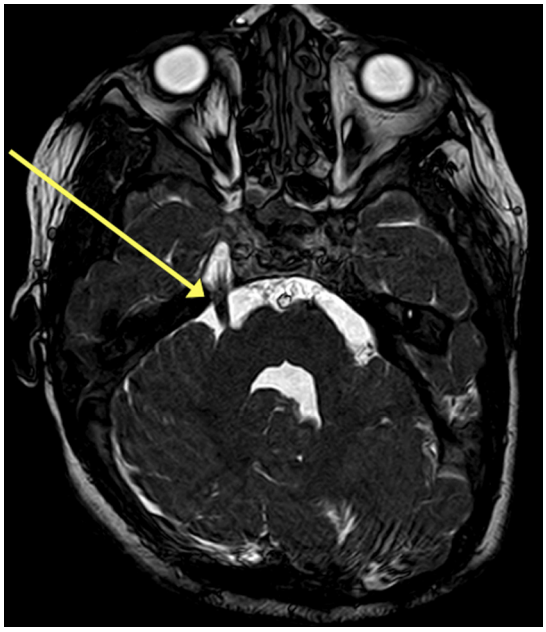

Fig. 1 – Magnetic resonance: left trigeminal nerve aplasia. The arrow points to the healthy contralateral trigeminal nerve.

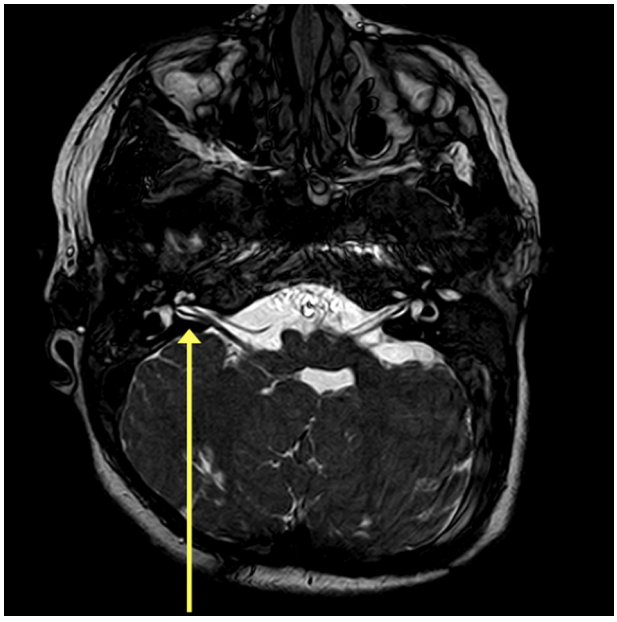

Fig. 2 – Magnetic resonance, hypoplasia of left facial and cochlear-vestibular nerves. The arrow points to the healthy contralateral structures.