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Case Report

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CORNEAL ANESTHESIA UNILATERAL: AN ATYPICAL CASE PRESENTATION

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ABSTRACT

Purpose: To report a case of unilateral corneal ulceration secondary to congenital agenesis of the trigeminal nerve associated with an arachnoid cyst of the left cerebellopontine angle. **Methods:** Case report. **Results:** We report the case of a 7-month-old girl referred to our department for a red eye with photophobia, without pain. Examination of the left eye revealed visual acuity limited to counting fingers at less than one meter, an abolished corneal reflex, absent corneal sensitivity, with infectious keratitis with hypopyon, and the fundus was unseen, leading to a normal ocular ultrasound. Ophthalmological examination of the Adelphe eye was normal. A notion of repetitive microtrauma of the left eye and self-mutilation of the left nose were detected during these follow-up visits. These symptoms were secondary to self-mutilation and associated with anesthesia of the left nose. Examination revealed the presence of homolateral left ear agenesis. Congenital agenesis of the trigeminal nerve associated with an arachnoid cyst of the left cerebellopontine angle pushes forward the homolateral acousticofacial nerve, which was confirmed by magnetic resonance imaging sections. The originality of our case lies in this particular association. To our knowledge, no case has been reported in the literature associating at the same time an arachnoid cyst of the cerebellopontine angle with an agenesis of the homolateral trigeminal nerve and of the homolateral ear with a corneal anesthesia, hence the particularity of our clinical case. **Conclusions:** Magnetic resonance imaging confirms congenital agenesis of the trigeminal nerve associated with an arachnoid cyst of the left cerebellopontine angle, a cause of infantile neurotrophic syndrome.

KEYWORDS: Child; neurotrophic keratitis; trigeminal agenesis; arachnoid cyst.

ABBREVIATIONS: CCA: Congenital corneal anesthesia MRI: Magnetic Resonance Imaging.

INTRODUCTION

Congenital corneal anesthesia (CCA) is a rare condition. It can be isolated or associated with neurological syndromes or polymalformations. The diagnosis must be suspected in front of any chronic corneal ulcer in the child. It is a condition that puts the visual prognosis at risk. Arachnoid cyst is a benign congenital formation containing cerebrospinal fluid. Most arachnoid cysts are small and asymptomatic and are located in the temporal fossa. Classically described complications are due to compression of adjacent structures.

OBSERVATION

We report the case of a seven-month-old infant, with no concept of consanguinity or medical history, suffering for a month from a unilateral corneal ulcer in the left eye. Slit lamp examination revealed a central corneal ulcer 6 mm by 5 mm in diameter with peri-lesion epithelial edema and hypopyon, in addition to conjunctival hyperemia and an altered tear film. However, the cornea did not show any foreign bodies. The study of corneal sensitivity found corneal anesthesia of the left eye **[figure 1],** the fundus was unseen, leading to a normal ocular ultrasound A notion of microtrauma to repetition of the left eye. self-mutilation of the left nose was detected at these follow-up visits. These symptoms were secondary to self-harm and associated with left nose anesthesia. The somatic examination revealed the presence of agenesis of the ipsilateral left ear**. [figure 2].**

Initial sampling of the cornea and conjunctiva with bacteriological and mycological studies was negative. Despite antibiotic drops and ointments (rifamycin and tobramycin), the result was negative. We decided to start an autologous serum after receiving the informed consent of the parents. The evolution after 1 month was marked by regression of the ulcer and edema and the disappearance of the hypopyon, but not the development of corneal opacity **[figure 3].**

As part of the etiological assessment of the corneal

anesthesia, a brain MRI in axial and coronal sections, in T2, diffusion and FIESTA sequences was performed, showing the presence of an arachnoid cyst of the left cerebellopontine angle, pushing forward the homolateral acousticofacial nerve bundle, and the absence of visualization of the left trigeminal nerve in its different portions: cisternal and at the level of Meckel's cavum. **[figure 4(A - E)].**

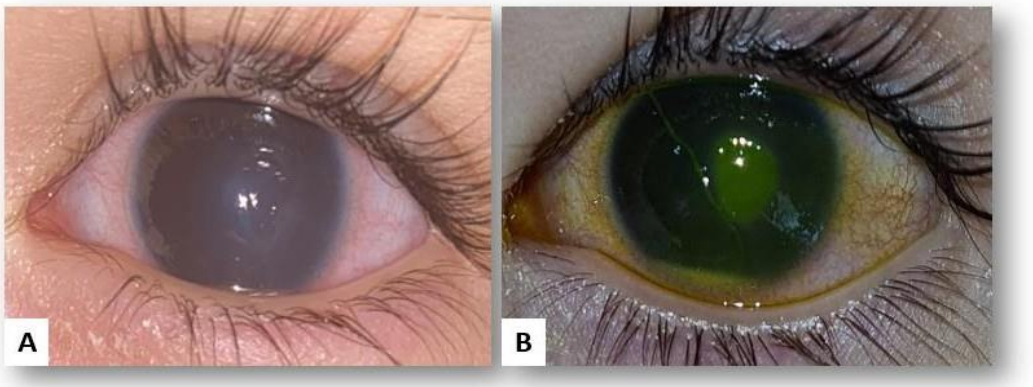

Figure 1(A and B): Biomicroscopic clinical aspect of the left eye showing a fluo+ulcer with pe rilesional edema and hypopyon.

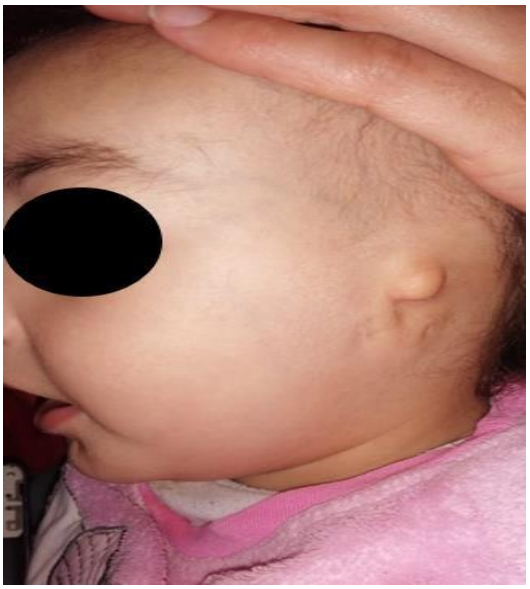

Figure 2: Photocolor of 7-month-old infant showing homolateral left ear agenesis.

Figure 3(A and B): Biomiscroscopic clinical appearance of the same eye after the start of treatment. A: Photocolor showing regression of the ulcer and edema. B: Color photo showing complete healing with appearance of central corneal opacity.

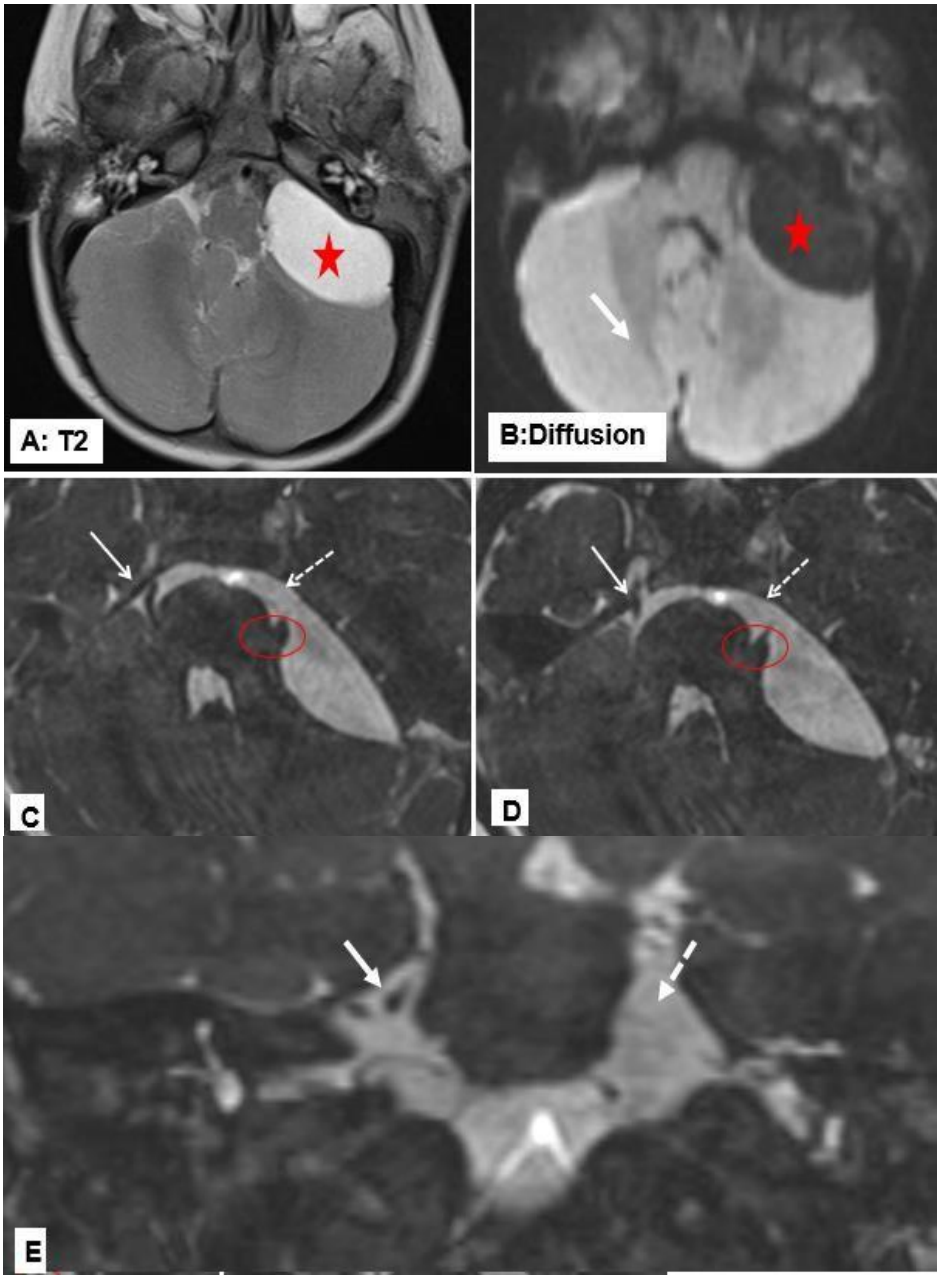

Figure 4(A-E): Brain MRI in axial (A-D) and coronal (E) sections, in T2, diffusion and FIESTA se- quences: an arachnoid cyst of the left cerebellar ponto angle (star), pushing forward the homo- lateral acoustic-facial nerve bundle. Normal right trigeminal nerve (white arrow).

DISCUSSION

Arachnoid cysts are malformations related to a developmental abnormality in which duplication of the primitive arachnoid leads to an intra-arachnoid collection of cerebrospinal fluid.

They constitute about 1% of intracranial masses. Cerebellopontine angle (CPA) arachnoid cysts are rare and often asymptomatic. The onset of symptoms and signs is usually due to compression of the brain, cranial nerves and obstruction of CSF circulation. The symptoms depend on the location and size of the cyst. The classically described symptoms of APC are headache, ataxia, seizures, and hemiplegia.

Although not very frequent, various ophthalmological manifestations can be the cause of the discovery of an arachnoid cyst: nystagmus, papillary hypoplasia, papillary edema, oculomotor paralysis.

The MRI appearance is quite typical Hyperintense lesion in T2 and hypointense in T1. On the diffusion sequence, the lesion appears hyposignal. In the pontocerebellar angle, the epidermoid cyst is one of the most frequently encountered congenital lesions. The latter has a hypersignal in diffusion.[1]

Cornea is the most innervated tissue in the human body through the ophthalmic branch of the trigeminal nerve. Corneal nerves are responsible for reflex lacrimation,

blinking, and the release of trophic factors, such as nerve growth factor, substance P, calcitonin, neuropeptide Y and acetylcholine. The blink and tear reflexes are important to protect the corneal surface from environmental factors and to supply essential nutrients and oxygen to the cornea. All of these factors promote epithelial cell proliferation, migration, adhesion, and differentiation and are necessary for the structural and functional integrity of the ocular surface.

Alteration of the corneal nerves leads to loss of corneal sensitivity and trophic functions, resulting in neurotrophic keratitis. The trigeminal nerve can be affected by systemic or ocular diseases, congenital or acquired, or iatrogenic injury.[2]

Congenital corneal anesthesia (CCA) is a rare condition that can be asymptomatic or manifest as neurotrophic keratitis. The first symptoms usually appear between the ages of 6 and 12 months. A chronic corneal ulcer is often the first sign.

The diagnosis of CCA is difficult and may be confused with other more common causes of neurotrophic keratitis, such as HSV or VZV keratitis, dry eye syndrome or recurrent epithelial erosions. It may be primary or secondary to a lesion involving the trigeminal nerve. It is essential to exclude acquired causes of corneal anesthesia before the diagnosis of CCA is made. Trigeminal nerve involvement may result from lesions involving the cerebellopontine angle, as was probably the case in our patient with her arachnoid cyst.[3]

CCA can be isolated or part of a syndrome such as Goldenhar syndrome (oculo-auriculovertebral dysplasia), Moebius syndrome (bilateral facial and abducens nerve palsy), VACTERL syndrome (vertebral, anal, cardiovascular, tracheo-esophageal, renal and limb), MURCS (Müller's duct aplasia, renal dysplasia, cervical vertebrae dysplasia), Riley-Day syndrome (familial dysautonomia).

The originality of our case lies in this particular association. To our knowledge, no case has been reported in the literature associating at the same time an arachnoid cyst of the cerebellopontine angle with an agenesis of the homolateral trigeminal nerve and of the homolateral ear with a corneal anesthesia, hence the particularity of our clinical case.[4]

The management of CCA is a therapeutic challenge, and progression to perforation is possible. The management of CCA is based on the prevention of accidental corneal injury resulting from corneal hypoesthesia and corneal self-trauma.

The authors agree that treatment with preservative-free artificial tears, autologous serum, and therapeutic contact lenses should be instituted to maintain ocular surface integrity, as well as antibiotics if needed. (Our patient progressed well on autologous serum).

Occlusion of the lacrimal point increases the retention of natural tears and improves the ocular surface. Surgical treatment is reserved for ulcers that do not respond to conservative treatment and includes: permanent tarsorrhaphy, conjunctival flap covering, or amniotic membrane grafting.[5,6]

Other treatments have been proposed in recent years: Regenerating agent (RGTA, Cacicol) is a matrix containing large polymers mimicking heparan sulfates that promotes healing.[7]

Corneal neurotization from the auricular or sural nerve.[8,9]

Recombinant Human Nerve Growth Factor (Cenegermin) has an effect on epithelial proliferation and the improvement of corneal sensory functions.[10] And more recently: allogeneic serum from the mother.[11] Transfixing corneal transplantation has a high risk of rejection. Corneal transplantation in these patients poses a very high risk, owing to poor wound healing and eventual vascularization that makes them liable for allograft rejection.[12]

Microtia is a relatively rare malformation characterized by characterized by hypoplasia of the auricle. Numerous reconstructive surgical techniques have been developed. Currently, the most widely used are autologous costal cartilage reconstruction and Medpor implant reconstruction. The authors present the indications for these two techniques, which can considerably improve quality of life for young patients. Reconstruction of the pinna using the Medpor can be performed on children as young as five years of age five years of age, and up to four years of age if the child is cooperative. Reconstructions using autologous costal cartilage require sufficient growth of the graft, which delays the procedure age of ten years. Morbidity is reduced in Medpor cases by limiting the number of anaesthesia procedures and avoiding costal cartilage ingrowth.[13-18]

CONCLUSION

CCA is a possible cause of chronic corneal ulceration in the pediatric population, which requires close monitoring by an ophthalmologist. A careful clinical examination is necessary to look for associated malformations.

It can lead to corneal scarring sequelae that may be responsible for severe amblyopia or even anatomical loss of the globe if not diagnosed in time.

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