Neurotrophic keratitis secondary to cerebral malformation☆

Queratitis neurotrófica secundaria a malformación cerebral

Dear Editor:

Neurotrophic keratitis is a degenerative disease of the cornea resulting from impaired trigeminal innervation. Prevalence is estimated at fewer than 5 cases per 10 000 population. Any noxa affecting the trigeminal nerve at any of its segments (from the nerve nucleus to its branches) may cause the disease. We present the case of a 7-year-old girl with a year-and-a-half long history of chronic right-eye keratitis. Local treatment with ocular lubricants yielded no improvement. A brain MRI performed at another hospital revealed no abnormalities. The ophthalmology department at our hospital detected complete corneal anaesthesia; the patient also reported increased sweating as well as discomfort when combing the hair on the right side of her head. Our patient, a Chinese-born girl, was adopted when she was 10 months old. Her year at school corresponds to her age. The physical examination revealed normal cranial nerve function except for mild right-sided asymmetrical wrinkling. The patient reported asymmetrical superficial tactile perception in her face; tactile perception was normal for the rest of the body. Tests for viral or bacterial infection yielded negative results. We requested a brain MRI scan with sequences centred on the posterior fossa; this revealed right trigeminal nerve and ipsilateral Meckel cave hypoplasia (Fig. 1).

Neurotrophic keratitis is a degenerative disease of the cornea resulting from trigeminal nerve alterations. The resulting lesions range from superficial punctate keratopathy to corneal ulcers or persistent epithelial defects which may lead to stromal melting and corneal perforation.[1,2] Any noxa affecting the trigeminal nerve may cause the disease. The most frequent causes of neurotrophic keratitis are herpes simplex virus infection, intracranial lesions, and surgery damaging the trigeminal nerve. Other causes include burns, wounds, corneal dystrophy, long-term use of local anaesthetics, or surgery of the anterior segment of the trigeminal nerve. A number of systemic disorders may also cause alterations in corneal sensitivity (multiple sclerosis, leprosy, etc.), familial dysautonomia, Goldenhar syndrome, Möbius syndrome, familial corneal hypoaesthesia, or congenital insensitivity to pain with anhidrosis.[1,3]

Physical examination may help locate the lesion. Presence of lagophthalmos suggests involvement of the sixth cranial nerve, whereas ptosis points to a lesion to the second cranial nerve. When the cause of the disease cannot be established clearly, an MRI scan centred on the posterior fossa should be performed to detect brainstem malformations.[2–4]

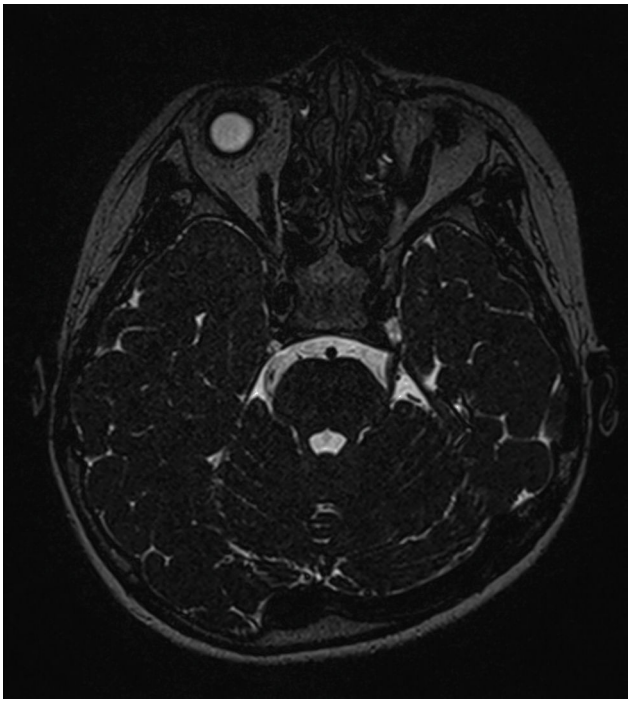

Figure 1 Axial sequence centred on the posterior fossa showing asymmetry in the sizes of the intracisternal portion of the trigeminal nerve and right Meckel cave.

There is no specific treatment for neurotrophic keratitis. Current treatment approaches focus on preventing corneal trauma and improving quality and transparency of the cornea. Different treatments can be used depending on lesion progression: local antibiotics, autologous serum, thymosin beta-4 eye drops, topical application of substance P, use of nerve growth factor, etc. Surgery is indicated only in severe cases. Amniotic membrane grafts seem to improve epithelialisation and reduce inflammation.[5–9] In conclusion, neurotrophic keratitis is an unusual condition with no specific treatment. It is rarely caused by anatomic lesions; however, brain malformations should be ruled out with neuroimaging studies of the brainstem.

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Conflicts of interest

The authors have no conflicts of interest to declare.

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Focal leptomeningeal uptake, a new radiological finding in pseudomigraine with pleocytosis☆

Captación leptomeníngea focal, un nuevo hallazgo radiológico en la seudomigraña con pleocitosis

Dear Editor:

Headache with neurological deficits and cerebrospinal fluid (CSF) lymphocytosis (HaNDL) is a syndrome characterised by[1] episodes of moderate or severe headache lasting a few hours,[2] cerebrospinal fluid with lymphocytic pleocytosis and normal neuroimaging results,[3] episodes of headache accompanied by transient neurological deficit, and[4] episodes of headache and neurological deficit recurring less than 3 months apart.[1] Although neuroimaging findings are usually normal,[2] some exceptions have been reported. We present the clinical case of a patient who met all diagnostic criteria for HaNDL syndrome and exhibited previously undescribed radiology findings.

Our patient was a 30-year-old man with no personal or family history of migraine who visited our department due to somnolence, mixed aphasia, right homonymous hemianopsia, and sensorimotor deficits in the right limbs. He had visited our department in the previous weeks reporting episodes of holocranial headache, sensory alterations, phonophobia, and photophobia; these symptoms had led to a diagnosis of migraine with sensory aura. Upon arriving at our hospital, our patient underwent a cranial CT scan and a neurosonological study, which yielded normal results. A few hours later he presented fever. A lumbar puncture performed after ruling out other possible focal infections disclosed clear CSF with 50 leukocytes (98% mononuclear), glucose levels of 69.1 mg/dL, and protein levels of 1.45 g/L.

A few days later, he experienced 2 episodes of holocranial headache associated with sensory deficits of the left upper limb which lasted a few hours. An additional lumbar puncture revealed clear CSF with 92 leukocytes (97% mononuclear), glucose levels of 61.7 mg/dL, and protein levels of 0.9 g/L. After that, he remained asymptomatic.

A brain MRI performed during his hospital stay revealed isolated leptomeningeal uptake in the posterior fossa (Fig. 1). Results from a series of blood and CSF screenings (including microbiology and cytology tests, a blood culture, autoimmune tests, and tumour marker tests) were normal. A thoracic-abdominal CT scan revealed no abnormalities.

A follow-up MRI scan performed 3 months later showed that leptomeningeal enhancement had resolved (Fig. 1). To date, 2 years later, our patient has experienced no further episodes.

HaNDL syndrome has been associated with focal slowing in EEG and focal alterations in blood flow as shown by different techniques (Doppler, CT perfusion, perfusion MRI, SPECT). The literature reports isolated cases of brain MRI abnormalities, including alterations in diffusion-weighted sequences at the level of the corpus callosum,[3] diffuse leptomeningeal enhancement,[4] or grey matter alterations associated with CSF enhancement in the temporal and occipital regions.[5] However, isolated leptomeningeal enhancement in the posterior fossa had never been described.

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