Short communication

Incomplete Horner’s syndrome as a presenting sign of fourth ventricle ependymoma

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ABSTRACT

Case report: The case of 44-year-old male patient with palpebral ptosis and trigeminal neuralgia as presenting sign of fourth ventricle ependymoma is reported. After surgical treatment, the patient developed a residual paresis of the sixth cranial nerve.

Discussion: Horner’s syndrome occurs due to an alteration of the sympathetic innervations of the eye and adnexa. Some tumors may be the cause, in our case an ependymoma of the fourth ventricle, which onset exceptionally with blepharoptosis and involvement of the ophthalmic division of trigeminal nerve, due to the proximity of these nerve fibers at the brainstem.

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Síndrome de Horner incompleto como signo de presentación de ependimoma del cuarto ventrículo

RESUMEN

Caso clínico: Varón de 44 años con ptosis palpebral y neuralgia del trigémino como signo de presentación de un ependimoma del cuarto ventrículo. Después del tratamiento quirúrgico desarrolló una paresia residual del sexto par.

Discusión: El síndrome de Horner aparece por la alteración de la inervación simpática del ojo y los anejos. Algunos tumores pueden ser la causa, en nuestro caso un ependimoma del cuarto ventrículo, que debutó de forma excepcional con ptosis palpebral y afectación de la división oftálmica del trigémino, debido a la proximidad de estas fibras nerviosas a nivel del troncocéfalo.

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Introduction

Ependymoma is a primary tumor of the central nervous system, of glial origin, exhibiting several degrees of histological malignity. Intracranial location is more frequent in children while the spinal cord is the location of choice in adults. Symptoms vary depending on location, size and degree of invasion and inflammation of the tumor.1–3 The most frequent ocular expressions are diplopia and diminished visual acuity, anisocoria, palpebral ptosis and papiledema (if it produces endocranial hypertension). The diagnosis is reached mainly through imaging techniques, with nuclear magnetic resonance (NMR) being the technique of choice although it is also possible to perform biopsies and spinal taps.

The treatment of choice is surgical exeresis although radiotherapy can be applied when the exeresis is not complete.

Clinic case report

A male, aged 44 years, visited the emergency service due to ptosis in the left eye (LE) of recent onset without other clinical signs. The patient is an active smoker and is on hypolipidemic medication.

An ophthalmological examination confirmed the presence of diminished palpebral groove in LE (8 mm), with adequate elevator muscle performance (16 mm). Exploration of both pupils under low lighting conditions did not evidence LE miosis. Upon the administration of a drop of apraclonidine hydrochloride 5 mg/ml (lopimax®) the ptosis disappeared. The ophthalmological examination did not produce additional relevant findings.

One month later the patient reported neuritic pain, radiating from the external edge of the LE toward the outer part of the ear, bipalpebral edema and congestion along the free edge which did not respond to corticoids topical treatment.

Chest X-ray and cerebral NMR were requested due to suspected incomplete Horner syndrome (Horner S.) (it was considered that the patient exhibited palpebral groove reductio due to dysfunction of the Müller muscle and the lower eyelid retractor, both with sympathetic innervation) and trigeminal neuralgia. The chest X-ray did not reveal alterations but the NMR exhibited a contrast-capturing tumoral lesion in the middle line of the posterior fossa at the level of the fourth ventricle base with hyper-capturing locations matching intratumor cysts. Ventricular size increases were associated. No metastasis was observed in the spinal cord (Fig. 1).

On the basis of location and radiological characteristics, the tumor was diagnosed as fourth ventricle ependymoma. The patient was referred to the neurosurgery service where it was decided to perform complete surgical exeresis with histological confirmation of the diagnostic (Fig. 2). A small cerebrospinal fluid (CSF) fistula without clinical repercussion was not extirpated and residual palsy of the fourth left cranial nerve with diplopia was observed, with complete resolution of the previous clinical expressions. The control resonances revealed a collection of CSF due to extradural fistula without tumor remains (Fig. 3). The evolution of said diplopia was followed up and, due to the persistence thereof 8 months after the tumor exeresis, it was decided to carry out a middle rectus retrinsertion (3 mm) in the right eye. Three years later, the patient remained free of symptoms.

Discussion

The usual presentation of Horner’s syndrome involves palpebral ptosis derived from Müller muscle palsy together with miosis and ipsilateral anhidrosis in preganglion cases. The lower eyelid is also involved with its retraction being diminished and giving the appearance of false enophthalmos. This could also be an incomplete Horner S. without some of the classical signs.

Eye drop tests can be applied to confirm the presence of the Horner S. and locate its cause. For many years 1% cocaine or hydroxyamphetamine tests have been applied but the difficulty of obtaining these products and its side effects have led to the use of more accessible drugs such as apraclonidine (lopimax®) which produces significant midriasis in the

Fig. 1 – Sagittal section in T1 of the brain resonance showing ependymoma in the fourth ventricle.

Fig. 2 – Histological section of the tumor with hematoxylin–eosin staining, 200×.
sympathetic nerve in the form of palpebral ptosis. Due to the compression exerted by the tumor on the spinal cord and encephalus, the sympathetic fibers descending from the hypothalamus could be involved. The fifth cranial pair can be affected by direct compression of the tumor or by the displacement of the nerve root over a blood vessel or the base of the cranium.5

The patient of this case exhibited subtle expressions but the successive appearance of 2 neurological signs such as incomplete Horner S. and trigeminal neuralgia together with a high degree of suspicion enabled early diagnosis and therapeutic surgical treatment with very few sequels for the patient.

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**Conflict of interests**

No conflict of interests has been declared by the authors.

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