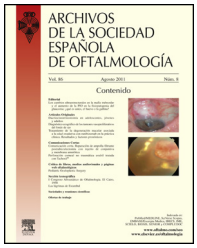
ARCHIVOS DE LA SOCIEDAD ESPAÑOLA DE OFTALMOLOGÍA

www.elsevier.es/oftalmologia



Short communication

Webino syndrome caused by meningovascular syphilis. A rare entity with an unexpected cause☆,☆☆

*M. Rodríguez Calvo de Mora**, *G. Rodríguez Moreno*, *M. España Contreras*

Servicio de Oftalmología, Hospital Regional Universitario Carlos Haya, Málaga, Spain

ARTICLE INFO

Article history:
Received 19 June 2012
Accepted 7 November 2012
Available online 5 August 2014

Keywords:
Wall-eyed bilateral internuclear ophthalmoplegia
Webino syndrome
Neurosyphilis
Meningovascular syphilis
Internuclear ophthalmoplegia

ABSTRACT

Case report: The patient is a 57-year-old obese and hypertensive male. His chief complaints were double vision and dizziness, with mild exodeviation in both eyes in primary gaze position in the ocular motility examination, but more predominant in the left eye. The exotropia was noticeably more evident on the attempted upgaze. On horizontal gaze, the abducting eye deviated fully, but the adducting eye did not cross the midline. Nystagmus in the abducting eye and convergence impairment were found. Pupil size and testing were normal. Ataxia and areflexia were also present. Bilateral internuclear ophthalmoplegia was suspected and imaging and laboratory tests were performed. The CAT scan showed a right occipital hypo-attenuated lesion. In the MRI scan, a mesencephalic subacute ischemic lesion was found, involving the medial rectus subnuclei. Blood and cerebrospinal fluid test for syphilis were positive.

Discussion: Bilateral internuclear ophthalmoplegia is a very uncommon–and difficult to diagnose–condition. In the reported case the lesion involved the medial rectus subnuclei. This fact could explain the exotropia in the primary gaze position, and supports that it is not possible to exclude the involvement of the medial rectus subnuclei in the webino syndrome. The rapid identification of the pathology contributed to the better prognosis of the patient.

Síndrome de webino secundario a sífilis meningovascular. Una entidad excepcional con una etiología inesperada

Palabras clave:
Oftalmoplejía internuclear bilateral
Síndrome de webino
Neurosífilis

RESUMEN

Caso clínico: Paciente varón de 57 años que refiere visión doble y mareo de aparición brusca. A la exploración oftalmológica se observó una exotropía evidente en posición primaria de la mirada, ausencia de aducción de ambos ojos, nistagmo en abducción e incapacidad para la convergencia. Ante la sospecha de oftalmoplejía internuclear (OIN) bilateral se

☆ Please cite this article as: Rodríguez Calvo de Mora M, Rodríguez Moreno G, España Contreras M. Síndrome de webino secundario a sífilis meningovascular. Una entidad excepcional con una etiología inesperada. Arch Soc Esp Oftalmol. 2014;89:199–202.

☆☆ This article was presented as a case report in the 87th Congress of the Spanish Society of Ophthalmology.

* *Corresponding author.*
E-mail address: marocalmo@gmail.com (M. Rodríguez Calvo de Mora).

Sífilis meningo-vascular
Oftalmoplejía internuclear

realizan pruebas de imagen y de laboratorio. La RMN craneal objetiva una lesión isquémica subaguda en el centro del mesencéfalo, afectando a los núcleos motores oculares comunes. Las pruebas para sífilis fueron positivas en sangre y líquido cefalorraquídeo.

Discusión: El síndrome de webino es muy infrecuente y de difícil diagnóstico. En el caso presentado, la lesión se encuentra perfectamente localizada en la zona media de la protuberancia, afectando a los núcleos motores oculares comunes. La rápida derivación del paciente y el establecimiento de tratamiento con penicilina posibilitaron la mejoría del cuadro.

Introduction

The gaze centers coordinate the action of motor nuclei for cooperating muscles to carry out symmetrical movements. The horizontal gaze center is the paramedian pontine reticular formation (PPRF), located in the protuberance close to the nucleus of the CN VI nerve pair. For horizontal gaze to take place the PPRF must activate the nucleus of CN VI which is comprised 2 types of neurons: motor neurons that innervate the ipsilateral rectus and internuclear neurons, whose axons cross the contralateral medial longitudinal fasciculus (MLF) and synapse in the subnucleus of the median rectus.[1]

In the event of MLF lesion, the connection of CN VI nucleus with the subnucleus of the contralateral median rectus does not take place. This produces and adduction deficit in the ipsilateral eye with normal gaze to that side because the opposite eye is able to adduct and the eye of the affected side exhibits normal abduction. It is frequent to observe disassociated nystagmus in horizontal sharp eye movements in abduction. This motor alteration is known as internuclear ophthalmoplegy (INO). It can be unilateral or bilateral and can be associated to vertical gaze palsy, vertical nystagmus and oblique deviation.[2]

The possible causes of INO include multiple sclerosis, myasthenia gravis, traumatisms, tumors, Wernicke's encephalopathy, infections such as syphilis, cryptococcosis and tuberculosis, the Arnold–Chiari malformation, occlusive vascular disease as well as some pharmaceutical drugs (barbiturates, lithium, and narcotics).

The *wall eyed bilateral internuclear ophthalmoplegia* (webino) syndrome is a particular type of bilateral INO with exotropia. It is a rare entity described by Lubow in 1971, characterized by horizontal gaze alteration exhibiting difficulty for adduction, exotropia in primary gaze position, nystagmus in the abducing eye and convergence inability.

The pathogeny of this peculiar form of INO is far from clear. It is known that most cases are caused by high brainstem injuries. The most accepted explanation for the cause is an injury in the midbrain[2] which bilaterally affects the MLF and both medium rectus subnuclei. Exotropia and convergence inability would be derived from the impairment of said subnuclei.

Other authors propose that the lesion is located at the pontine and doubt that the involvement of the rectus muscle subnuclei is an essential condition. Even though the majority of reported cases derive from mesencephalic injuries, some publications report that the syndrome is caused by isolated pontine injuries.[3,4]

The webino syndrome etiology is similar to that of INO: vascular causes are more frequent, followed by multiple sclerosis and traumatism. A post-surgery webino syndrome has also been reported.[5]

Clinical case

A diabetic, hypertense and obese male patient, aged 57, smoker and with moderate drinking habit, in treatment with metformin and enalapril, visited the emergency ophthalmology section referring double horizontal vision with dizziness and instability, stable from onset. General examination revealed ataxic gait and areflexia.

Ophthalmological exploration produced a corrected visual acuity (BCVA) of 0.7 in the right eye and difficult 0.8 in the left eye. Anterior pole exploration and funduscopy with midriasis did not produce significant findings other than slight lens sclerosis. Intraocular pressure was within normal limits. Pupil reflexes were normal. Eye movement exploration revealed severe exotropia in primary gaze position, absence of adduction in both eyes, nystagmus abduction in both eyes and absence of convergence (Fig. 1). Simple and alternate cover test revealed alternating exotropia. Bielchowsky maneuver evidenced discrete skew. In levoversion, major vertical nystagmus (*downbeat*) was evidenced.

Due to suspected bilateral INO, imaging and lab tests were carried out. Cranial CAT casually revealed right occipital hypoattenuated injury. Cranial NMR in T2 flair mode evidenced a shiny pointed lesion affecting the subnuclei of ocular motor nerves, with ischemic and subacute appearance (Fig. 2). The ischemic origin of the lesion was confirmed by the black color of the affected area in the ADC map (Fig. 2). Similarly, the NMR diffusion image showed involvement of the same area (Fig. 3).

A range of exhaustive tests was carried out with samples of blood, urine and cerebrospinal fluid (CSF). Serology revealed positive total antibodies for syphilis and RPR (1/64). Pn CSF, FTA (1/2) and VDRL (1/1) were positive. Additional findings were pleiocitosis and protein increase. Accordingly the condition was diagnosed as *meningovascular syphilitic meningitis*.

The patient was admitted for supervision, treatment and additional studies. Treatment was established with IV penicillin (24,000,000 daily units during 10 days), with partial remission of motor ocular alterations, predominantly in the left side. Three months after diagnosis, the condition subsisted partially with persistent left side INO. With one year of evolution, the patient exhibits slight and residual persistence of left side ophthalmoplegy.