POSTSURGICAL WEBINO, A NEW FORM OF THIS SYNDROME

SÍNDROME DE WEBINO POSTQUIRÚRGICO, UNA NUEVA FORMA DE ESTE SÍNDROME

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ABSTRACT

Case report: A 20 year-old woman was submitted to resection of an ependymoma of the fourth ventricle. After surgery the patient began to refer diplopia and oscillopsia. WEBINO syndrome (wall-eyed bilateral internuclear ophthalmoplegia), vertical nystagmus and skew deviation were diagnosed.

Discussion: WEBINO syndrome is considered a special form of bilateral internuclear ophthalmoplegia. It is characterized by bilateral absence of adduction, nystagmic abduction of both eyes, convergence deficiency and frequently exotropia. Ischemic, demyelinating and infectious etiologies have been described, but to our knowledge this is the first report of a postsurgical form of this syndrome (Arch Soc Esp Oftalmol 2009; 84: 407-410).

Key words: Wall-eyed bilateral internuclear ophthalmoplegia, WEBINO, ependymoma.

RESUMEN

Caso clínico: Una mujer de 20 años de edad fue sometida a resección de un ependimoma del cuarto ventrículo. Después de la cirugía la paciente comenzó a sufrir diplopia y osciloscopia, siendo diagnosticada de síndrome de WEBINO (wall-eyed bilateral internuclear ophthalmoplegia), nistagmus vertical y desviación oblicua (skew).

Discusión: El síndrome de WEBINO es considerado una forma especial de oftalmoplejia internuclear bilateral. Se caracteriza por la ausencia de aducción y nistagmus en abducción en ambos ojos, ausencia de convergencia y con frecuencia exotropia. Hasta el momento se han descrito etiologías isquémicas, desmielinizantes e infecciosas. Sin embargo probablemente este es el primer caso de síndrome de WEBINO de etiología posquirúrgica publicado.

Palabras clave: Oftalmoplejía internuclear bilateral, WEBINO, ependimoma.

CLINICAL CASE

A 20-year-old woman without relevant personal history was admitted for operation due to posterior fossa tumor adjacent to the fourth ventricle. The patient began with hydrocephalia, for which a ventricle-peritoneal valve had been implanted for controlling intracranial pressure. Two weeks later she was submitted to a second operation in order to resect the tumor. In the post period she exhibited...
multiple comblinocular diplopia, oscillopsia, deviation of the lips, dysphagia and weakness in all limbs. The physical exploration revealed preserved superior functions, paresia of adduction in both eyes together with nystagmus in abduction in both eyes (fig. 1), isochoric and normo-reactive pupils and tetraparesia 4/5 in arms and 3/5 in lakes, lower left facial paresia and left deviation of the tongue with ipsilateral atrophy of hemi-tongue due to paresia of the left hypoglose. The anatomic-pathological studies showed that the tumor is an ependymome of the fourth ventricle, WHO degree 2. The post surgery cranial magnetic resonance evidenced remains of the tumor with hemorrhagic component in the region over the fourth ventricle as well as signal changes associated to restriction of diffusion in bilateral frontoparietal region is compatible with brain infarcts of border territories in relation to low expenditure episodes (fig. 2). This cortical ischemic damage in border territories explained the tetraparesia.

In the course of six months, tetraparesia and the ophthalmological condition have exhibited a favorable evolution (fig. 1). At the time of writing this article, exotropia has disappeared and convergence has improved, changing the conditions to conventional bilateral internuclear ophthalmoplegia.

**DISCUSSION**

The oculomotor alterations exhibited by the patient correspond to a clinical condition described only occasionally in the literature and known under the acronym of WEBINO (wall-eyed bilateral internuclear ophthalmoplegia).
This peculiar form of internuclear bilateral ophthalmoplegia, the pathogenic of which is far from clear (1), is characterized by the association of adduction limitation in both eyes accompanied by nystagmus in the abducting eye, exotropia in primary gaze position (contrary to what occurs in patients with internuclear ophthalmoplegia were even in bilateral cases exhibit orthophoria) and incapacity for convergence.

In this case the patient also exhibited hypertropia of the LE (oblique or skew deviation) and vertical nystagmus. Both expressions appear frequently associated to internuclear ophthalmoplegia (2). In addition, oblique deviation has a certain localization value because usually the hypertropic eye is the ipsilateral eye to the damaged medial longitudinal fascicule (2).

To date, we haven’t found in the literature any revision about the syndrome and all publications referring to this mysterious entity correspond to small series of isolated cases. The etiology matches that of internuclear ophthalmoplegia (2), the majority of cases being of demyelinizing or ischemic causes (3). There seems to be a certain ethnic influence and the majority of cases published in Japan had ischemia causes while in the European countries and the United States the demyelinizing etiology predominates. Some post-traumatic cases have been described (4). Another exceptional etiology is that of infections caused by cryptococosis (4) or tuberculosis (5).

From the pathogenic viewpoint it is known that most cases are produced by high lesions in the trunk of the encephalus. Therefore, the most extended hypothesis states that it is due to the combined lesion of the medial longitudinal fascicule and the two sub-nuclei of the middle rectals. This added lesion of the sub-nuclei of the middle rectals would explain the frequently associated exotropy and the incapacity of convergence. In this sense, it would represent the high variance of the 1.5 syndrome involving a simultaneous lesion of the medial longitudinal fascicule and the nucleus of the sixth cranial pair. However, some authors have revised in full detail small series of this syndrome and have concluded that said sub-nuclei need not necessarily be affected (1). The lesion which account for the insufficient convergence could also be located at the level of the cerebellum or the internal capsule (4).

Considering the poor general condition of the patient, treatment with botulin toxin or surgery was dismissed. After six months of evolution the patient exhibited a spontaneous improvement, with a disappearance of diplopia and the convergence insufficiency. The clinical condition turned into a conventional internuclear ophthalmoplegia. Against what could be expected initially, a partial improvement of the clinical condition has taken place.

REFERENCES