TERSON'S SYNDROME ASSOCIATED WITH ARNOLD-CHIARI I MALFORMATION

SÍNDROME DE TERSON EN MALFORMACIÓN DE ARNOLD CHIARI I

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ABSTRACT

Case report: We present a patient with bilateral Terson’s syndrome after brainstem surgical decompression as treatment for her Arnold-Chiari I malformation. We have studied and compared the progression of preretinal hemorrhage after Nd:YAG laser treatment of the right eye and observation of the left, and also the formation of an epiretinal membrane in the left eye.

Discussion: Treatment of Terson’s syndrome is based on observation or surgery according to different criteria. Hyaloidectomy with YAG laser produces good results in selected cases (Arch Soc Esp Oftalmol 2007; 82: 113-116).

Key words: Terson syndrome, vitreous hemorrhage, subhyaloid hemorrhage, Arnold-Chiari Malformation, epiretinal membrane.

INTRODUCTION

Terson’s Syndrome (TS) is defined as a vitreous hemorrhage followed by any type of intracranial bleeding, due to an intracranial hypertension transmitted through the space of the optic nerve sheath (NOS) (1). The Arnold-Chiari malformation is characterized by a caudal displacement of the posterior fossa structures through the foramen magnum.

RESUMEN

Caso clínico: Se presenta una paciente con Síndrome de Terson (ST) bilateral tras descompresión quirúrgica medular como tratamiento de su malformación de Arnold Chiari tipo I. Se estudia y compara la progresión de sus hemorragias prerretinianas tras tratamiento con láser Nd:YAG en su ojo derecho y observación en el izquierdo, formándose en éste una membrana epirretiniana.

Discusión: El tratamiento del ST se basa en observación y/o cirugía según ciertas pautas. La hialoidectomía YAG también consigue buenos resultados en casos seleccionados.

Palabras clave: Síndrome de Terson, hemorragia vítrea, hemorragia subhialoidea, Malformación de Arnold-Chiari, membrana epirretiniana.
CASE REPORT

A 45 year-old woman referred from the neurosurgery service due to sudden loss of bilateral vision after a ventricular-peritoneal derivation surgery of a temporal subdural hygrome and cranial occipital abscess. Said condition was the result of a subacute postsurgical complication. Fifteen days earlier she had been intervened for preventive medullar decompression due to the Arnold-Chiari I malformation (figs. 1,2).

The patient had a clinical condition going 16 years back, with migraines of slight intensity, instability, rotatory dizziness, self-restricted vomits of variable duration and slight vertical nystagmus a year ago. The latter condition led to the suspicion and diagnosis of the congenital malformation because the rest of symptoms related to the ulcerating colitis she suffered since years ago.

The visual acuity (VA) in the first exploration was movement of hands in both eyes. The pupillar reactions were of 2+ without afferent pupillar relative defect, exhibiting a high frequency and mean amplitude descending nystagmus. The eye fundus showed retrohyaloid premacular and vitreous bilateral hemorrhage, confirmed with optical coherence tomography (OCT) and ocular echography (figs. 3, 4). All these findings were compatible with TS.

A posterior hyaloidectomy was performed in the right eye (RE) with previous instillation of anesthetic eye drops and use of Nd laser:YAG 15 pulses (4-7 range mJ/pulse) in the most inferior part of the hematoma. The posterior hyaloid broke and drained immediately to the vitreous cavity (fig. 5). It was decided to maintain an expectant attitude regarding the left eye (LE).

Visual acuity after one year, following several revisions with remarkable improvements, was of 0.6 in RE and 0.1 in the LE. The OCT revealed a premacula epiretinal membrane of recent appearance in the LE (fig. 6). The frequency of the nystag-
mus went down and the amplitude became slight. The bilateral hemovitreous continued to be in slow and progressive reabsorption. The patient rejected any ocular surgery, being satisfied with the visual result attained.

**DISCUSSION**

The treatment of the Arnold-Chiari malformation is surgical, by means of the decompression of the cranial-vertebral union (2).

The early complications of this technique are very rare and include meningitis, hematoma, infection and subdural hygroma. Our patient underwent simultaneously the two last complications, which were solved with derivation techniques.

Some complications of the lumbar and ventriculoperitoneal derivations have been described, such as intracranial hemorrhage after the passage of the catheter through the brain parenchyma and tonsilar herniation to with intracranial hypertension (2). The latter caused the premacular hemorrhage in the case patient, as described above.

In TS, intracranial hypertension is transmitted within the NO sheath because it dilates, compressing and obstructing in turn the retinochoroidal anastomosis. All this leads to a noticeable reduction of the ocular venous drainage with subsequent hypertension and retinal venous hemorrhage (1).

The treatment options of TS include observation, pars plana vitrectomy (3) or administration of Nd: YAG laser to drain hematoma to the vitreous cavity (4). The indication of each one depends on the characteristics of the patient.

The observation can be considered as the first indication according to the following factors: unilaterality, extension of the vitreous opacity, signs of spontaneous reabsorption, age, occupation, visual necessities, psychological and neurological state, family role and risk of new surgery.

The vitreous hemorrhage in the TS is solved very slowly in a year and the average of visual improvement oscillates between 20/300 and 20/30. The hemovitreous can persist up to 6 years (3).

The complications of a persistent preretinal hemorrhage are: epiretinal membrane (from 25 to 70% according to different studies), retinal hole, disruption of the retinal pigmentary epithelium and toxic retinal effects of the hemoglobin (3).

Three months can be a reasonable period for the observation and determination of the vitreous clearing. Unless contraindicated by the general conditions (as in our case) vitrectomy should be performed.

In selected cases Nd: YAG laser can be used for the photodisruption of the hematoma, but the surgeon and patient must be aware of its possible complications, among which we emphasize the macular hole, retina detachment and formation of epiretinal membrane (5).

In our patient, the latter option of treatment was adopted in the RE, since she was a good collaborator and would not require general anesthesia. The therapy was carried out safely and without subsequent complications. However, this was not the case
for the LE, which developed an epiretinal membrane after one year of observation.

REFERENCES