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Case Report

Sudden Loss of Vision as a Result of Idiopathic Intracranial Hypertension: A Case Report

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Abstract

Introduction: Pseudotumor cerebri syndrome is a condition characterized by an increase in intracranial pressure without demonstrable cause, without evidence of ventricular dilatation and with papilledema.

Case Report: 37-year-old male patient with a history of anemia without known cause and Idiopathic Intracranial Hypertension for 2 years, which was treated conservatively. A year and a half later, he returned to the clinic again because he reported an abrupt loss of vision in only 3 days without headache. Complementary studies were performed and showed an increase in intracranial pressure and an increase in the transverse diameter of both optic nerves in imaging studies, so it was decided to perform emergency surgical treatment.

Discussion: Microsurgical fenestration of the optic nerve sheath by means of a medial or lateral transscleral orbitotomy and lumbo-peritoneal CSF derivation were the techniques proposed to solve the increase in intracranial pressure. Lumboperitoneal shunting was finally decided as definitive treatment due to the bilateral atrophy of the papilla presented by this patient. Seven days after surgery, MRI studies of the orbit show normal recovery of the transverse diameter of the optic nerves. Although the patient does not improve the visual loss, the progression to amaurosis is stopped.

Conclusions: Ideopathic intracranial hypertension is a disease that although its highest incidence is in the female and obese population, it can occur in any patient. The follow-up of asymptomatic patients in neuroophthalmology consultation prevents residiva, identifying papilledema from its incipient form, which favors treatment in its early stages and avoids its progression to optic atrophy.

***Keywords:** cerebral pseudotumor, idiopathic intracranial hypertension, papilledema, optic nerve sheath fenestrations, lumboperitoneal shunt.*

Introduction

Pseudotumor cerebri syndrome is a condition characterized by an increase in intracranial pressure without a demonstrable cause, without evidence of ventricular dilatation and with papilledema. In most cases it occurs in young and obese women, without a clear explanation [(1)]. The noxa was described by Quinke in 1897, but it was not until 1914 that this term began to be used. Shortly thereafter, Foley suggested using the term benign endocranial hypertension as it seemed to indicate that this increase in intracranial pressure had a much more favorable course than those produced by infectious, or tumor causes. However, due to the great morbidity of the visual pathway suffered by these patients, the term "benign" fell into disuse; other more appropriate terms are primary cerebral pseudotumor or idiopathic endocranial hypertension and secondary cerebral pseudotumor [(2,3,4,5,6)].

The problem with the term is that there is a direct relationship between stenosis, bilateral or not, of the transverse sinuses and the syndrome. Many of the patients once diagnosed with idiopathic endocranial hypertension were later shown to have transverse sinus stenosis. Patients who present with transverse sinus stenosis, venous sinus thrombosis, or in the rare cases where a relationship between drug use and increased intracranial pressure is demonstrated, should fall into the category of secondary pseudotumor cerebri [(1)].

Pseudotumor cerebri occurs in children and young adults. Ninety percent of these are due to idiopathic increased intracranial pressure. The estimated incidence in the United States of America is 0.9/100000 in the general population, with a female-to-male ratio of 8:1 (7,8). In the female population the incidence increases to 3.5/10000 in ages between 20 and 44 years. In those with an increase of 10% of their ideal weight the incidence rises to 13/10000 and to 19/10000 in those with 20% above their ideal weight. Rapid weight gain is considered a risk factor for developing cerebral pseudotumor in both sexes [1,7,9,10].

We present a young, thin, male patient, with no risk factors, who developed idiopathic endocranial hypertension with abrupt loss of vision.

Clinical Case

A 37-year-old male patient with a history of anemia with no known cause for which he was under hematological study, who 2 years ago began with oppressive headache, preferably in the morning, relieving during the day and sometimes waking him up in the early morning, with a month of evolution. He went to Neurology consultation because he began to notice progressive loss of vision in the last week, finding bilateral chronic papilledema on physical examination. For these reasons, imaging studies were indicated and when these did not reveal expansive pathological processes or ventricular dilatation, as well as thrombosis of venous sinuses or stenosis of transverse sinuses, Idiopathic Endocranial Hypertension (IIH) was suspected and a Lumbar Puncture (LP) with manometry was performed, confirming elevated figures in the Cerebrospinal Fluid (CRL) pressure. Serial LP evacuations and treatment with acetazolamide were performed and the patient showed absolute improvement of the picture with recovery of vision, so he started to be followed by Neuroophthalmology and Neurology consultation. With the resurgence of the Covid-19 pandemic and the confinement, this patient stopped the follow-up by consultation for more than a year and a half; and returns again because he refers that in only 3 days, he has had an abrupt loss of vision, without previous headache or abrupt weight gain. Reasons for which the patient is sent to Neurosurgery consultation.

Physical examination

General Physical Examination.

Normolinear patient with difficulty walking because he requires his companion to guide him so as not to trip over objects.

Weight: 65kg

Physical Examination by Systems

SN: Patient vigilant, oriented in time, space and person, who responds to questioning with clear and coherent language. No meningeal signs or motor defect. Glasgow scale: 15 points.

Cranial IIpair:

Best corrected visual acuity: Counting vision.

Color vision: Hishihara test: 2/23.

Fundus: Pale-yellowish papilla corresponding with bilateral papillary atrophy is observed in both eyes.

Imaging studies:

Computerized axial tomography of the skull and orbits (CTS): CTS is performed in 1mm slices where normal brain parenchyma is observed, with centered midline and without alterations in the ventricular system. In orbits, bilateral thickening of both optic nerves is observed in more than 7mm in its transverse diameter. (Fig.1)

Brain and orbits magnetic resonance study (MRI): There was no evidence of thrombosis of venous sinus or stenosis of transverses sinus. Increase in more than 7mm the optic transverse diameter.

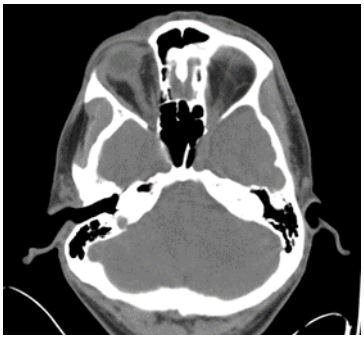

Fig.1: *Increased transverse diameter of the optic nerve*

Optical coherence tomography (OCT): adequate thickness in the nerve fiber layer is observed in the RNFL Thickness Average technique, however ganglion cells cannot be scanned for measurement in the macula because the patient does not achieve the required focus due to his lack of vision.

Invasive studies

Lumbar puncture with manometry: 32mmHg.

Discussion

Because this was a patient diagnosed with an IIH with abrupt loss of vision, it was decided to perform emergency surgery to decrease intracranial pressure and thus relieve CSF compression on the optic nerves. Microsurgical fenestration of the optic nerve sheath by means of a medial or lateral transscleral orbitotomy and lumbo-peritoneal CSF shunting were the techniques proposed to resolve the increased intracranial pressure. Considering that this is a male patient, without abrupt weight gain and of normolinear constitution, that the abrupt loss of vision was not preceded by headache, microsurgical fenestration of the optic nerve sheath by means of a medial or lateral transscleral orbitotomy [11,12,13] was an attractive option for this patient because it is an efficient technique that requires the fenestration of a single optic nerve with the consequent improvement of both optic nerves. In a retrospective study carried out by Neel S Vaidya et al. where 104 patients diagnosed with cerebral pseudotumor and who underwent microsurgical fenestration of the optic nerve sheath were analyzed, a resolution of papilledema was found in 76% in the first week, with no loss of vision 6 months after surgery [14]. In another study evaluating the safety of the method when performed for the second and third time in patients with IIH, it is shown that it continues to be effective in the resolution of papilledema even after up to three attempts [15].

However, lumbo-peritoneal CSF shunting is a simpler and equally effective method for resolving papilledema and is still the first choice of treatment for many neurosurgeons today. Although both procedures have a relatively low morbidity, the most frequent complications associated with lumbo-peritoneal shunting are the descent of the cerebellar tonsils and the palliative nature of the system, since it must be revised every time it malfunctions. A study publicated by Georgios Niotakis in 2013, where is analyzed every patient with diagnosis of IIH between 2005 and 2010 in a single center, found it that 15 patient treated with lumboperitoneal shunt required at lest a shunt revision.[16]. In a study comparing both methods, no significant differences were found between them in terms of safety and resolution of papilledema. In another study published in Indian J Ophthalmol, which reviews the subject, it is concluded that the first treatment option for IIH continues to be controversial and that microsurgical fenestration of the optic nerve sheath is recommended for those patients with loss of visual acuity without headache, recommending lumbo-peritoneal shunting for the remaining group [17,18]. Although it is true that this patient had IIH with visual loss without headache, the physical examination and OCT had shown that he had bilateral optic atrophy, so the possibility of visual recovery was null. It was then decided to perform a lumbo-peritoneal shunt in order to stop the progression of the disease and prevent the patient from reaching bilateral amaurosis. Seven days after surgery, the patient had no worsening of the condition, so an MRI of the orbits was performed, where optic nerves with adequate diameters of less than 4mm (Fig.2) were observed and the effectiveness of the treatment was confirmed.

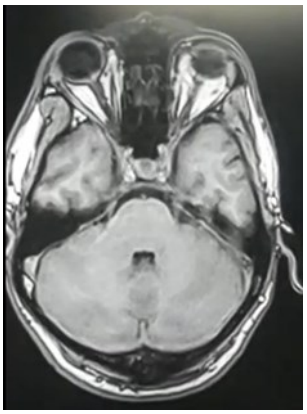

Fig. 2: *Normal transverse diameter of the optic nerve*

Conclusions

IIH is a disease that although its highest incidence is in the female and obese population, it can occur in any patient, so it requires the evaluation of a multidisciplinary team that includes neurologists, neuro-ophthalmologists and neurosurgeons. Due to the high morbidity of vision, the treatment, conservative or surgical, should always pursue the objective of total resolution of papilledema. The follow-up of asymptomatic patients by neuro-ophthalmology consultation prevents residiva, identifying papilledema from its incipient form, which favors treatment in its early stages and avoids its progression to optic atrophy.

Conflicts of Interest: None

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