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CLINICAL CASE

Severe ocular damage secondary to a pseudotumor cerebri

Daño ocular severo secundario a un seudotumor cerebri

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Abstract

Introduction: *Pseudotumor cerebri causes symptoms of intracranial hypertension and visual alterations secondary to damage to the optic nerve and the retina.* **Case Report:** *Hispanic 11-year-old male, obese. Presents to ER with recurrent visual alterations and vomit. Besides arterial hypertension and fundoscopy with hemorrhages, exudate, papilledema and silver-wire vessels. With a normal cranium tomography and a cerebrospinal fluid pressure of 38 cm H2O, management was started for pseudotumor cerebri. Visual recovery was limited due to severe damage to the retina and the optic nerve.* **Conclusions:** *In pseudotumor cerebri, visual damage can be permanent if proper treatment is not initiated early. With extensive retinopathy, malignant hypertension must be ruled out.*

Key words: *Pseudotumor cerebri. Malignant hypertension. Retinopathy. Papilledema.*

Resumen

Introducción: *El seudotumor cerebri ocasiona síntomas de hipertensión craneal y alteraciones visuales secundarias a daño del nervio óptico y retina.* **Presentación del caso:** *Masculino, raza hispana, 11 años, obeso. Acude por cefalea, vómito recurrente y alteraciones visuales. Además de hipertensión arterial y fundoscopia con hemorragias presenta exudados, papiledema y arteriolas en cable de plata. Con una tomografía de cráneo normal y raquimanometría de 38 cmH2O, se inició manejo para seudotumor cerebri. La recuperación de la visión fue limitada por daño severo en retina y nervio óptico.* **Conclusiones:** *En el seudotumor cerebral, el daño de la visión puede ser permanente si no se instaura un tratamiento oportunamente. Ante una retinopatía extensa, se debe descartar una hipertensión arterial maligna.*

Palabras clave: *Seudotumor cerebri. Hipertensión maligna. Retinopatía. Papiledema.*

Introduction

Pseudotumor cerebri, or idiopathic or benign intracranial hypertension (ICH), is defined as the elevation of intracranial pressure without an apparent cause, that is, not due to mass effect, cerebrospinal fluid (CSF) absorption failure or acute brain inflammation; however, it is said that these terms are erroneous since in many occasions causal factors are found or the ocular manifestations can be very severe and irreversible[1]. Pseudotumor cerebri is a condition that is observed more frequently in adult women, obese, of childbearing potential, and may be associated with the intake of substances such as vitamin A, antibiotics (tetracyclines),

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hormonal medications (oral contraceptives, danazol, levonorgestrel) or steroids and their abrupt suppression, among others. The appearance under 20 years of age is unusual and unlike adults, in prepubertal children, it can occur without a sex predilection; however, in 30-43% of obese children and only as age increases, obesity constitutes a risk factor for pseudotumor cerebri, being reported in up to 80-90% of adolescents aged 15-17 years with obesity[2,3].

The typical symptomatology is that of intracranial hypertension, with headache, tinnitus, nausea, vomiting and visual alterations1 such as visual field defects, increased blind spot size, diplopia due to paresis of the VI cranial nerve, transient ischemia, photopsia, retrobulbar pain, papilledema, up to irreversible atrophy of the optic nerve; and in 5% of the cases blindness that occurs in fulminant idiopathic ICH, a very rare entity. To make the diagnosis (see Table 1), it is necessary to rule out organic causes of elevated intracranial pressure, have a normal brain computed tomography or magnetic resonance imaging (MRI), lack of focal neurologic signs and intracranial pressure greater than 250 mmH2O, that even in children ranges between 250 and 470 mmH2O, with normal composition of the CSF[4,5].

Ocular fundus damage secondary to idiopathic ICH comprises optic disc swelling with specific findings according to the severity of retinal hemorrhages and papilledema (Table 2)[3,6].

Among the differential diagnoses to be ruled out are the causes of papilledema accompanied by intracranial hypertension and visual alterations, such as posterior fossa tumors, intracranial hemorrhage, venous sinus thrombosis, and, in this review, we emphasize malignant hypertension; a rare disease in the pediatric age that defines a series of changes in the retina such as hemorrhages and exudates, together with systemic arterial hypertension (SAH). Although the prevalence of hypertension in children is of 3-5%, there are few studies about the prevalence of hypertensive retinopathy in children, estimated between 8% and 50%7. Ocular damage in malignant SAH is described in Table 3.

We present the case of a patient with severe retinal damage due to pseudotumor cerebri.

Table 1. Dandy-Smith criteria

Signs and symptoms of intracranial hypertension
Lack of focal neurologic signs, except for VI nerve paresis
Normal neuroimaging tests (small ventricles and "empty sella" appearance may be present)
Increased CSF pressure (above 200-250 mmH20) but with normal composition

Table 2. Fundus findings in pseudotumor cerebri

Typical papilledema findings
Blurred disc margins
Absence of venous pulsation
Retinal veins distension
Optic disc protrusion
Peripapillary hemorrhages
Peripapillary exudates

Table 3. Hypertensive retinopathy classification

Mild	Generalized arteriolar narrowing, arteriovenous nicking, silver wire arterioles
Moderate	Retinal hemorrhages (dot or flame-shaped), microaneurysms, cotton-wool spots, hard exudates
Severe	Moderate retinopathy signs plus optic disc swelling

Case presentation

An 11-year-old male patient with diagnosis of severe obesity, weighing 91 kg (P99) with a height of 1.52 cm (P50-75) and a body mass index of 39 (P > 97). With a family history of essential hypertension and type 2 diabetes mellitus. He attended the ophthalmology department for a 5-week history of headache described as holocranial, pulsatile and of variable intensity, sometimes disabling, as well as a sudden visual loss in the right eye 2 weeks before, having the same sudden visual loss later in the left eye, a week prior to admission. His relatives did not seek medical attention before due to family problems. The patient has been living in the school residence away from his parents for the last 6 months.

He went to the emergency room a month and a half before due to headache, ataxia and vertigo with a blood pressure (BP) of 120/90 (percentile P 88/P99). Acute otitis media was diagnosed, and he was discharged. In the ophthalmology department the diagnosis was of hypertensive retinopathy with a BP of 140/90 (P99/P99), so he was referred for admission to the pediatrics department. In his initial assessment he no longer referred headache, nausea or vomiting and the BP was normal; however, 15 hours after admission, the patient presented a BP of 140/100 mmHg (P99 / > 99). The other vital signs were normal, with a Glasgow score of 15. He referred blurred vision and "shadows" in the right eye, and in the left eye his visual acuity was of light perception. Fundoscopy showed retinal hemorrhages

and papilledema, corroborated by digital camera examination where extensive bilateral hemorrhages, microaneurysms, cotton-wool spots and hard exudates, grade IV papilledema, macular star, and silver wire arterioles were observed in both eyes (Fig. 1). The rest of the physical exploration was normal.

For the diagnostic approach of the patient, a computed tomography with and without contrast was performed, which was normal. Blood tests showed white and red cell counts within normal ranges with a blood glucose of 105 mg/dL in fed state. Urinary, renal and liver enzymes tests were normal. A lipid profile was not quantified. A lumbar puncture was performed, due to suspicion of pseudotumor cerebri, which reported an opening pressure of 38 cmH2O and CSF of normal composition. With this data, a diagnosis of pseudotumor cerebri was made and treatment was initiated with acetazolamide 45 mg/day and topiramate 50 mg/day, showing a slight improvement of the symptoms. During his hospital stay the BP remained in a range of normal to high with values between 110-120/70-90 mmHg; after 5 days he was discharged with the same treatment, referring a slight improvement in vision; however, he was not evaluated by the ophthalmology department before discharge.

Two months after discharge, an improvement in visual acuity was reported. The patient was able to see blurred objects and the visual acuity measured with a Rosenbaum card was 20/200 in the left eye and 20/800 in the right eye. The BP was in the normal range in this visit. At 9 months, with a weight of 85 kg, his visual acuity was 20/100 in the right eye and 20/400 in the left eye, without a refractory error. And on fundoscopy, the retina showed fibrosis secondary to the inflammatory event, and both optic nerves were pale.

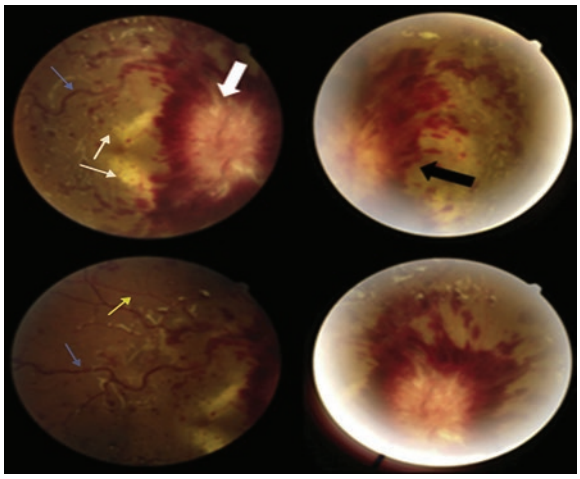

Figure 1. Patient's ocular fundus. Flame and punctate hemorrhages are observed (black arrow), macular edema (thick white arrow), silver wire arterioles, arteriolar constriction (yellow arrow), venous congestion and tortuosity (blue arrow) and exudates (thin white arrows).

Discussion

We present a patient with severe visual alterations secondary to pseudotumor cerebri with hypertension. The only direct association of pseudotumor cerebri with hypertension is when the patient has hypertension secondary to obesity, but hypertension has not been described as part of the clinical signs. It has only been reported that previously hypertensive patients have a worse visual prognosis, and that obese patients have a higher probability of recurrence[6]. In this case, hypertension was attributed to the patient's obesity; however, other possible causes of hypertension and its severity were not studied.

Pseudotumor cerebri may show fundus changes as those seen in malignant hypertension, nevertheless, it is unusual that there is extensive retinal damage as in this case. On the other hand, hypertensive retinopathy in children is usually mild[7], contrary to the findings in this patient, who would be classified as a severe case. Therefore, malignant hypertension should be considered in the diagnostic approach of patients with BP elevation and severe retinal damage, as in this case. Considering these two options was a challenge for the service. First, other differential diagnoses had to be excluded, especially those related to mechanical ICH such as tumors, intracranial hemorrhage and venous sinus thrombosis. Posterior fossa tumors should be ruled out due to the visual symptoms in our patient. Posterior fossa syndrome is characterized by headache, nausea, vomiting and papilledema (10-15%), visual disturbances and uncoordinated gait. MRI should be performed as a more sensitive imaging test to diagnose common tumors in childhood such as medulloblastoma, astrocytoma, ependymoma or craniopharyngioma[8]. Another differential diagnosis to consider is subarachnoid hemorrhage that causes ICH symptoms due to inflammation and fibrosis. Some examples are aneurysms, trauma or rarely an arteriovenous malformation (10% are in the posterior fossa). An aneurysm can rupture due to hypertension, causing ICH symptoms due to acute hydrocephalus (blood is released to the CSF), which can cause sentinel headache, starting 6-20 days before the

event itself in 30-50% of patients. A hemorrhage can be excluded by computed tomography[9]. Finally, venous sinus thrombosis, a condition more common in neonates and children than in adults, causes ICH due to thrombosis or occlusion, which causes a decrease in CSF absorption. The signs and symptoms are variable, but most cases are subacute (50% of cases), occurring between 2 and 30 days and presenting with localized and progressive headache in 89% of cases, which increases with the Valsalva maneuver. ICH manifestations are more common in the chronic presentation (1-6 months). This condition must be ruled out with MRI or computed tomography, as well as a computed venography or venous resonance[10].

In our patient, an MRI was not performed due to lack of equipment in our institution; however, these entities were excluded by computed tomography, a somewhat less sensitive tool, but which, together with signs and symptoms and the patient×s evolution, allowed us to discard those possibilities.

Regarding pseudotumor cerebri and malignant hypertension, both diseases are unusual in the pediatric age, so there is little literature about them.[11] In our patient, unlike that reported in the literature[12] in cases of pseudotumor cerebri, the recovery of visual capacity was limited, with optic nerve atrophy[13]. We consider this outcome was due to late diagnosis and treatment. Since permanent visual loss is the most serious pseudotumor cerebri or malignant hypertension sequel, it is necessary to establish a rapid and accurate diagnosis in these conditions to prevent progression[14].

Conclusion

The diagnosis of pseudotumor cerebri should be considered among several diseases that show ICH signs and symptoms. It is a diagnosis of exclusion, and an ophthalmologic assessment is essential because permanent visual loss is the most severe sequel. When there is severe retinal damage, malignant hypertension should be excluded. A timely diagnosis is crucial for an adequate response to treatment and to avoid visual sequelae.

Ethical disclosures

Protection of human and animal subjects. The authors declare that the procedures followed were in accordance with the regulations of the relevant clinical research ethics committee and with those of the Code of Ethics of the World Medical Association (Declaration of Helsinki).

Confidentiality of data. The authors declare that they have followed the protocols of their work center on the publication of patient data.

Right to privacy and informed consent. The authors have obtained the written informed consent of the patients or subjects mentioned in the article. The corresponding author is in possession of this document.

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Conflicts of interest

The authors declare no conflict of interests.

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