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Importance of fulminant idiopathic intracranial hypertension: case report

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Introduction

Idiopathic intracranial hypertension (IIH) is a neurological condition predominantly affecting women, with obesity being the main risk factor. Symptoms include migraine, orthostatic headaches, transient visual obscurations, visual blurring, pulsatile tinnitus and diplopia. The prognosis worsens with faster progression. Understanding the clinical presentation and managing it efficiently are crucial, although a minority of patients present with the fulminant form of the disease.

Objective

To report a case of fulminant IIH.

Case Report

A 33-year-old female patient with no comorbidities and a BMI of 29.5 was admitted to a neurology hospital in Curitiba, PR, due to progressive decrease in visual acuity (VA), bilateral tinnitus and headaches that worsened when lying down. Physical examination revealed bilateral papilledema with VA of 20/200 in the left eye and 20/50 in the right eye. Cranial CT without relevant findings. Lumbar puncture with an opening pressure >50 cm H2O and no inflammatory signs. Pharmacological treatment with acetazolamide was initiated, gradually increasing to two tablets every 6 hours, later adjusted to every 8 hours due to side effects. Topiramate 25 mg twice daily was added, but visual complaints persisted. MRI and MRA revealed bilateral sigmoid sinus stenosis. After ruling out secondary structural and metabolic causes, a diagnosis of IIH was considered. During hospitalization, acetazolamide was suspended because of electrolyte disturbances (metabolic acidosis and hypokalemia), with only topiramate being continued. Due to worsening visual acuity after 8 days of hospitalization, a right frontal ventriculoperitoneal shunt was performed with neuronavigation assistance. After recovery, patient discharged without headache or tinnitus complaints, with severe VA 20/260 (left eye) and moderate VA 20/100 (right eye). At a follow-up outpatient, the patient reported resolution of headache, relief of left pulsatile tinnitus, and partial improvement of bilateral visual changes, although right visual impairment was more pronounced and bilateral papilledema persisted, despite regression. For follow-up, brain MRI, computerized visual field testing, optical coherence tomography, fluorescein angiography, and neuro-ophthalmologic consultation were requested.

Conclusion

Due to the urgency of the fulminant form of IIH, it is crucial to monitor the rapid and progressive evolution of visual loss, characteristic of the pathology, in order to minimize neurological sequelae.

