Fulminant Idiopathic Intracranial Hypertension Managed with Ventriculoperitoneal Shunting and Bilateral Optic Nerve Sheath Fenestration

Hipertensão Intracraniana Idiopática Fulminante Tratada com Derivação Ventriculo-peritoneal e Fenestração da Bainha do Nervo Ótico Bilateral

Mariana Portela [1], André Sobral-Pinho [2], Tiago Lorga [3], João Costa [1]

[1] Ophthalmology Department, Centro Hospitalar Universitário de Lisboa Ocidental, Lisbon, Portugal
[2] Neurology Department, Centro Hospitalar Universitário de Lisboa Ocidental, Lisbon, Portugal
[3] Neuroradiology Department, Centro Hospitalar Universitário de Lisboa Ocidental, Lisbon, Portugal

Recebido/Received: 2022-10-15 | **Aceite/Accepted:** 2023-05-23 | **Publicado/Published:** 2023-06-26

DOI: https://doi.org/10.48560/rspo.28242

ABSTRACT

Fulminant idiopathic intracranial hypertension (IIH) is a rare form of IIH with rapid and aggressive vision loss. Urgent management is required to prevent blindness. We present the case of a 28-year-old woman with new symptoms of transient visual obscurations, headache and pulsatile tinnitus with findings of severe bilateral optic disc edema that developed devastating visual loss. Blood tests, brain imaging and cerebral spinal fluid analysis excluded secondary causes. A lumbar puncture revealed intracranial hypertension and magnetic resonance imaging findings supported it. A ventriculoperitoneal shunt was placed, but due to continued vision deterioration, an urgent bilateral optic nerve sheath fenestration was performed with slight vision improvement. This case highlights the relevance of early recognition of this entity and the demand for prompt consideration of surgical management to prevent further visual loss and blindness. It also illustrates the importance of multidisciplinary work and low vision rehabilitation for a young, visually impaired adult.

KEYWORDS: Decompression, Surgical; Hypertension, Malignant; Intracranial Hypertension/surgery; Optic Nerve/surgery; Papilledema; Pseudotumor Cerebri; Ventriculoperitoneal Shunt.

RESUMO

A hipertensão intracraniana idiopática (HII) fulminante é uma forma rara de HII que cursa com perda visual rápida e agressiva. A abordagem urgente é necessária para prevenir a cegueira. Apresentamos o caso de uma mulher de 28 anos com sintomas de obscurações visuais transitórias, cefaleia e acufenos pulsáteis, e edema do disco ótico bilateral, que desenvolveu deterioração visu-

al grave. Estudos analíticos, exames de imagem cerebral e avaliação do líquido cefalorraquidiano excluíram causas secundárias. A punção lombar mostrou aumento da pressão intracraniana com achados corroborados pela ressonância magnética cerebral. Foi colocada derivação ventrículo-peritoneal, mas por manter deterioração visual foi realizada fenestração da bainha do nervo ótico bilateral urgente com discreta melhoria visual. Este caso realça a importância do reconhecimento precoce desta entidade e a necessidade de considerar intervenção cirúrgica célere para prevenir perda visual e cegueira. Mostra-se ainda a relevância do trabalho multidisciplinar e de reabilitação visual numa doente jovem com baixa visão.

PALAVRAS-CHAVE: Derivação Ventriculoperitoneal; Descompressão Cirúrgica; Hipertensão Intracraniana/cirurgia; Hipertensão Maligna; Nervo Óptico/cirurgia; Papiledema; Pseudotumor Cerebral.

INTRODUCTION

Idiopathic intracranial hypertension (IIH) is characterized by high intracranial pressure of unknown cause, after careful exclusion of secondary causes. It was first described by Quincke *et al* in 1897 and later coined pseudotumor cerebri in 1904. In 1955 Foley *et al* named the disease benign intracranial hypertension but due to later reports of severe and permanent visual loss and intractable headache, the term "benign" was dropped.[1,2] IIH is estimated to affect 1 to 2 per 100 000 people per year, and its incidence is higher in overweight women between the ages of 15 to 44 years.[3] It usually presents with transient visual obscurations or visual loss related to postural changes, pulse-synchronous tinnitus, frontal or retro-orbital headaches, nausea and vomiting.[4] It is characterized by bilateral papilledema on fundoscopy and an enlarged blind spot or peripheral vision loss in visual field testing.[5] Magnetic resonance imaging (MRI) and magnetic resonance venography of the brain may suggest intracranial hypertension (empty sella, flattening of the posterior aspect of the globe or transverse sinus stenosis) but are usually performed to exclude secondary causes of increased intracranial pressure.[4] Lumbar puncture reveals an elevated cerebral spinal fluid (CSF) opening pressure above the normal level of 25 cm H2O with normal CSF composition.[6] Prompt intervention is essential, and the main goals of treatment are vision preservation and symptom control. Medical treatment is usually enough and includes weight reduction and oral acetazolamide.[7]

Fulminant IIH is a rare (2,9%) form of IIH characterized by severe vision deterioration within 4 weeks of symptom onset and continued vision loss over days. Urgent surgical intervention is usually mandatory to improve visual prognosis.[8,9] The reason why some patients develop a severe rapid evolution in a characteristically subacute to chronic disease is still unknown.[9] Current surgical options include ventriculoperitoneal (VP) or lumboperitoneal (LP) shunts and optic nerve sheath fenestration (ONSF).[10] CSF diversion is mainly performed in cases of severe headache or continued vision loss despite maximal medical treatment. ONSF is usually indicated for papilledema-related vision loss. Refractory patients with dural venous sinus stenosis may benefit from venous sinus stenting.[10]

IIH is usually a self-limited disorder, but in some cases, it can recur or even become chronic.[10] In 5% to 10% of patients, it may lead to blindness.[11]

CASE REPORT

A retrospective review of clinical records was performed to assess clinical and treatment outcomes. Informed consent to publish this case and its images was obtained from the patient.

A 28-year-old woman presented to the emergency room with a 4-day history of intense occipital headaches associated with transient visual obscurations and bilateral pulsatile tinnitus. Her medical history was significant for obesity, migraine, hormonal contraceptive use, and systemic arterial hypertension during her last pregnancy. On examination, her blood pressure measured 172/124 mmHg. Neurological examination and a brain computed tomography (CT) were unremarkable. Best corrected visual acuity (BCVA) was 0.2 in the right eye (OD) and 0.5 in the left eye (OS), and color vision was affected in both eyes (Ishihara plates of 8/13 in the OD and 11/13 in the OS). Ocular motility, tonometry, and anterior segment examination were normal. Pupil light reflexes were present but sluggish, and there was no evident relative afferent pupillary defect. A dilated retinal examination showed bilateral Frisén grade 4 disc edema, bilateral retinal vein tortuosity with congestion and flame-shaped hemorrhages in the OS (Fig. 1).

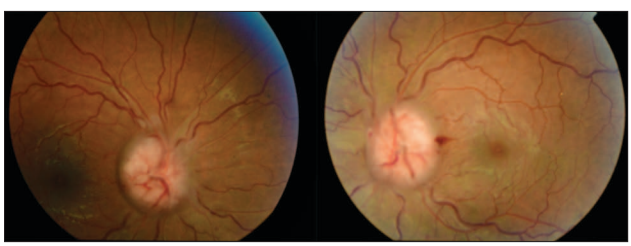


Figure 1. Retinographies of the right and left eyes showing bilateral optic disc edema with retinal vein tortuosity and flame shaped hemorrhages.

Sublingual captopril (25 mg) was administered, and the blood pressure decreased to 149/95 mmHg. A complete blood count, clotting, renal and liver profiles, brain computed tomography (CT) and CT venography were all unremarkable. Venous sinus thrombosis and intracranial mass lesions were therefore excluded. A subsequent lumbar puncture measured an opening CSF pressure higher than 40 cm H2O with normal CSF composition. A diagnosis of IIH was admitted, and oral acetazolamide 500 mg was started twice daily. On day 2 of admission, the patient complained of aggravated visual loss and headache. Her blood pressure levels remained under 140/90 mmHg and malignant systemic hypertension was also excluded. A brain MRI demonstrated an intrasellar meningocele and a bilateral optic nerve head protrusion, suggesting severe intracranial hypertension (Fig. 2).

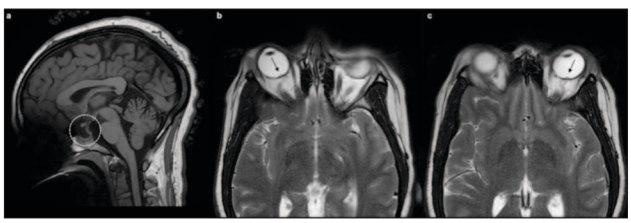


Figure 2. Brain MRI demonstrating signs of intracranial hypertension. 2a: Intrasellar meningocele (circle) | 2b and 2c: Bilateral intraocular optic nerve head protrusion (arrows).

Considering the serious decline in visual function, fulminant IIH was considered, and the patient was submitted to a surgical intervention for the placement of a VP programmable shunt at the lowest pressure. On day 3 after admission, a slight improvement regarding headache and bilateral vision was reported by the patient, but 2 days later, she complained of a new worsening of these symptoms despite a functioning shunt. The BCVA was light perception in the OD and no light perception in the OS. A temporizing CSF lumbar drainage was implanted with a slight improvement of headache and vision, and a bilateral ONSF was programmed for the next day. Bilateral ONSF was performed, on day 6 after admission, through a medial orbital approach and starting with the OD.

A medial limbal conjunctival peritomy was performed and the medial rectus muscle was isolated and secured with a double armed 6–0 vicryl suture. The muscle was detached from the globe, leaving a small remnant of tendon to which a traction suture was attached. The globe was then retracted laterally and the optic nerve sheath was exposed with the aid of small malleable retractors. Using an ophthalmic 15º side port blade, and with caution to avoid any blood vessel on the surface of the nerve, a small window was cut on the sheath 2 mm posterior to the globe, allowing CSF to egress into the orbit. No intraoperative complications were observed.

The CSF lumbar drainage was removed on the same day. On day 8 after admission, her BCVA improved to counting fingers in the OD and hand motion in the OS and on the following days to 0.2 bilaterally despite a very narrow bilateral visual field demonstrated on confrontational testing. Her headache and tinnitus also improved, and a significant reduction of the disc edema was observed bilaterally on fundoscopy. The patient was discharged, and regular neuro-ophthalmology, neurosurgery, psychiatry, and low-vision follow-up visits were provided. Visual rehabilitation was started 4 months after diagnosis, and +1D lenses for near vision with occlusion of the non-dominant eye were prescribed to improve reading speed. Practice with reverse telescopes, visual field expanders, and visual scanning training was also initiated. She was referred to a vision rehabilitation center for orientation and mobility training. One year later, the BCVA had improved to 0.8 bilaterally, but with very narrow visual fields (almost 0º in the OD and less than 5º in the OS), strongly affecting her vision quality (Fig. 3).

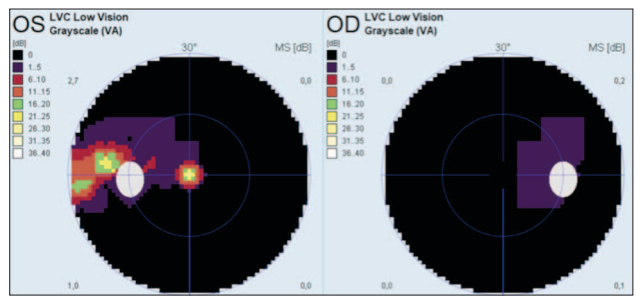


Figure 3. Standardized automated perimetry with the low vision program demonstrating severe bilateral constriction of the visual fields one year after the diagnosis of fulminant IIH.

The fundoscopy and optical coherence tomography (OCT) demonstrated severe bilateral optic nerve atrophy (Fig. 4).

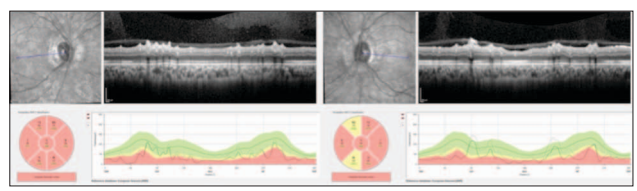


Figure 4. OCT showing severe bilateral optic disc atrophy one year after the diagnosis of fulminant IIH.

Currently, the patient is able to walk around the house unaided and perform some usual daily activities, such as cooking and cleaning, on her own. However, despite the substantial visual improvement observed and the new orientation skills gained, the patient is still unable to work and is currently unemployed. She is also on psychiatry and psychology appointments due to depression.

DISCUSSION

We report the case of a 28-year-old obese woman diagnosed with fulminant IIH that had severe acute visual

acuity deterioration and refractory headache and that was submitted to 3 consecutive surgical interventions, namely a VP shunt, a temporizing lumbar drain and finally a bilateral ONSF.

Initially, malignant systemic hypertension diagnosis was also considered a possible cause of bilateral optic neuropathy and headache. Malignant hypertension is defined as diastolic blood pressure greater than or equal to 120 mmHg or systolic blood pressure greater than or equal to 180 mmHg and is a known cause of such findings.[6,12] Because the blood pressure decreased to reasonable levels after medication and did not rise again during follow-up, and despite the increased arterial pressure during pregnancy, there was no evidence of other hypertensive values in subsequent post-partum medical appointments, we assumed fulminant IIH as the primary diagnosis. Furthermore, there were no secondary causes for hypertension after appropriate investigation. However, the coexistence of systemic hypertension cannot be ignored and may have had implications on the worse prognosis. A similar case, although with higher blood pressure values (220/140 mmHg), has been described in the literature.[13]

Fulminant IIH is characterized by severe and rapidly progressive vision loss and requires urgent and aggressive treatment to preserve visual function, sometimes including temporizing procedures such as lumbar drainages.[9] The highly incapacitating visual course of our case led us to consider such strategies. Typical cases of IIH, however, are characterized by less deterioration of visual acuity and simply require medical and behavioral treatment. Surgical management is reserved for refractory patients.[10]

In fact, the most significant recent meta-analysis studying surgical management of refractory IIH proposes that venous sinus stenting should be regarded as the first-line treatment.[14] We did consider implanting a transverse sinus stent in our patient, but the risk of a Labbé vein thrombosis was considered too high to perform this procedure in this specific case safely.

A CSF diversion procedure is one of the most common surgical procedures for fulminant IIH and has been reported to relieve all symptoms, including vision loss. There is, however, some controversy regarding stability and duration of effect, even with functioning shunts.[10] Despite a functional shunt, our patient´s visual function continued to decline.

In the same meta-analysis, which consisted of 818 patients and 1365 eyes, papilledema, visual field defects, and headaches improved in 90.5%, 65.2%, and 49.3% of cases, respectively, after ONSF. In a large study reporting the results of 158 ONSFs performed in 86 patients, visual acuity stabilized or improved in 94%, and visual fields stabilized or improved in 88%.[15] Furthermore, there are also some reports on cases with aggravating visual loss that required an ONSF despite a functioning shunt.[16,17]

In an attempt to stop the severe decline of of our patient's visual function, we decided to perform an ONSF 4 days after the VP shunt placement, with mild success.

Vision rehabilitation and low-vision visits must not be forgotten and may constitute one of the most critical steps in long-term follow-up. A recent systematic review concluded that vision rehabilitation does have a beneficial effect on vision-related quality of life, as was also the case of our patient.[18]

In addition, we also point out that a multidisciplinary approach, contemplating neurosurgery, neurology, neuroradiology, neuro-ophthalmology, psychiatry, and low vision teams was essential in diagnosis, management, and follow-up.

CONCLUSION:

- Fulminant IIH is characterized by severe and rapidly progressive vision loss and requires urgent and aggressive treatment to preserve visual function and improve headache.
- Current surgical options include ventriculoperitoneal or lumboperitoneal shunts, optic nerve sheath fenestration and venous sinus stenting.
- Although rare, some cases require more than one surgical procedure, namely an optic nerve sheath fenestration despite a functioning shunt.
- Multidisciplinary work and low vision rehabilitation are of paramount importance.

CONTRIBUTORSHIP STATEMENT / DECLARAÇÃO DE CONTRIBUIÇÃO:

All authors provided critical feedback and helped shape the research, analysis, and manuscript. MP and ASP wrote the first draft of the manuscript. MP, ASP, JT and TL contributed to the material preparation, data collection and analysis. PA and JC oversaw the overall direction and planning of the study.

RESPONSABILIDADES ÉTICAS

Conflitos de Interesse: Os autores declaram a inexistência de conflitos de interesse na realização do presente trabalho.

Fontes de Financiamento: Não existiram fontes externas de financiamento para a realização deste artigo.

Confidencialidade dos Dados: Os autores declaram ter seguido os protocolos da sua instituição acerca da publicação dos dados de doentes.

Consentimento: Consentimento do doente para publicação obtido.

Proveniência e Revisão por Pares: Não comissionado; revisão externa por pares.

ETHICAL DISCLOSURES

Conflicts of Interest: The authors have no conflicts of interest to declare.

Financing Support: This work has not received any contribution, grant or scholarship.

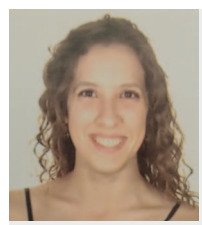
Confidentiality of Data: The authors declare that they have followed the protocols of their work center on the publication of data from patients.

Patient Consent: Consent for publication was obtained.

Provenance and Peer Review: Not commissioned; externally peer reviewed.

REFERENCES

1. Corbett JJ, Savino PJ, Thompson HS, Kansu T, Schatz NJ, Orr LS, et al. Visual loss in pseudotumor cerebri. Follow-up of 57 patients from five to 41 years and a profile of 14 patients with permanent severe visual loss. Arch Neurol. 1982; 39:461–74. doi: 10.1001/archneur.1982.00510200003001
2. Wall M, Hart WM Jr, Burde RM. Visual field defects in idiopathic intracranial hypertension (pseudotumor cerebri). Am J Ophthalmol. 1983; 96:654–69. doi: 10.1016/s0002-9394(14)73425-7
3. Lee A, Wall M. Idiopathic intracranial hypertension (pseudotumor cerebri): Epidemiology and pathogenesis. [accessed Oct 2022] Available from: http://www.uptodate.com/ contents/idiopathic-intracranial-hypertension-pseudotumorcerebri- epidemiology-and-pathogenesis.
4. Lee A, Wall M. Idiopathic intracranial hypertension (pseudotumor cerebri): clinical features and diagnosis. [accessed Oct 2022] Available from: http://www.uptodate.com/ contents/ idiopathic-intracranial-hypertension-pseudotumorcerebri-clinical-features-and-diagnosis.
5. Jensen RH, Radojicic A, Yri H. The diagnosis and management of idiopathic intracranial hypertension and the associated headache. Ther Adv Neurol Disord. 2016;9:317-26. doi: 10.1177/1756285616635987.
6. Headache Classification Committee of the International Headache Society (IHS) The International Classification of Headache Disorders, 3rd edition. Cephalalgia. 2018;38:1-211. doi: 10.1177/0333102417738202.
7. Smith SV, Friedman DI. The Idiopathic Intracranial Hypertension Treatment Trial: A Review of the Outcomes. Headache. 2017;57:1303-10. doi: 10.1111/head.13144.
8. Bouffard MA. Fulminant idiopathic intracranial hypertension. Curr Neurol Neurosci Rep. 2020;20:8. doi: 10.1007/s11910-020-1026-8.
9. Thambisetty M, Lavin PJ, Newman NJ, Biousse V. Fulminant idiopathic intracranial hypertension. Neurology. 2007;68:229–32. doi: 10.1212/01.wnl.0000251312.19452.ec.
10. Lee A, Wall M. Idiopathic intracranial hypertension (pseudotumor cerebri): prognosis and treatment. [accessed Oct 2022] Available from: http://www. uptodate.com/contents/ idiopathic-intracranial-hypertensionpseudotumor-cerebri-prognosis-and-treatment.
11. Greenberg M. The Handbook of Neurosurgery. 8th ed. New York: Thieme Publishers; 2016.
12. Mollan SP, Davies B, Silver NC, Shaw S, Mallucci CL, Wakerley BR, et al. Idiopathic intracranial hypertension: consensus guidelines on management. J Neurol Neurosurg Psychiatry. 2018;89:1088-100. doi: 10.1136/jnnp-2017-317440.
13. Abbasi HN, Brady AJ, Cooper SA. Fulminant idiopathic intracranial hypertension with malignant systemic hypertension-a case report. Neuroophthalmology. 2013;37:120-3.
14. Kalyvas A, Neromyliotis E, Koutsarnakis C, Komaitis S, Drosos E, Skandalakis GP, et al. A systematic review of surgical treatments of idiopathic intracranial hypertension (IIH). Neurosurg Rev. 2021;44:773-92. doi: 10.1007/s10143-020-01288-1.
15. Banta JT, Farris BK. Pseudotumor cerebri and optic nerve sheath decompression. Ophthalmology. 2000;107:1907-12. doi: 10.1016/s0161-6420(00)00340-7.
16. Kelman SE, Sergott RC, Cioffi GA, Savino PJ, Bosley TM, Elman MJ. Modified optic nerve decompression in patients with functioning lumboperitoneal shunts and progressive visual loss. Ophthalmology. 1991;98:1449-53. doi: 10.1016/s0161-6420(91)32113-4.
17. Sergott RC, Savino PJ, Bosley TM. Modified optic nerve sheath decompression provides long-term visual improvement for pseudotumor cerebri. Arch Ophthalmol. 1988;106:1384-90. doi: 10.1001/archopht.1988.01060140548021.
18. van Nispen RM, Virgili G, Hoeben M, Langelaan M, Klevering J, Keunen JE, et al. Low vision rehabilitation for better quality of life in visually impaired adults. Cochrane Database Syst Rev. 2020;1:CD006543. doi: 10.1002/14651858.CD006543.pub2.



Corresponding Author/
Autor Correspondente:

Mariana Portela
R. da Junqueira 126,
1349-019 Lisboa, Portugal
E-mail: marianasportela@gmail.com

ORCID: 0000-0002-5352-4292