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Severe visual loss from concurrent fulminant idiopathic intracranial hypertension and malignant arterial hypertension: Prompt suspicion matters

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ABSTRACT

Purpose: To report a case series of 4 patients with poor visual outcomes from concurrent fulminant idiopathic intracranial hypertension (IIH) and malignant arterial hypertension with bilateral optic disc edema. The diagnosis of fulminant IIH was delayed given the bilateral optic disc edema was attributed initially to hypertensive optic neuropathy.

Observation: All 4 patients (3 males, 3 African Americans, mean BMI 27.6 kg/m^2 (range 19.5–36 kg/m^2) presented to the emergency department with bilateral vision loss, optic disc edema, and blood pressure (BP) of greater than 180/120. The patients were treated initially to control BP and the optic disc edema was either attributed to the hypertension or the ophthalmic examination was not performed. The patients were subsequently diagnosed with IIH with Brain MRI, MR venogram, and lumber puncture (mean cerebrospinal fluid (CSF) opening pressure 42 cm, range 40–43 cm). The mean time from presentation to diagnosis of IIH was 3.2 months (range 1–6 months). The final visual acuity ranged from 20/400 to hand motions in the better eye and count fingers to hand motions in the worse eye despite bilateral optic nerve sheath fenestrations (3 patients), ventriculoperitoneal shunts (3 patients), and treatments with acetazolamide (3 patients) and furosemide (1 patient).

Conclusion: Our case series underscores the need to promptly include IIH in the differential diagnosis in patients with bilateral optic disc edema including patients with malignant hypertension, particularly in those experiencing progressive visual loss, regardless of gender or BMI. Prompt work-up with brain MRI with contrast and MR or CT venogram to detect neuroimaging signs of intracranial hypertension followed by a lumbar puncture with CSF opening pressure are essential to initiate rapid treatment of fulminant IIH to avoid poor outcome.

1. Introduction

Idiopathic intracranial hypertension (IIH) is characterized by elevated intracranial pressure (ICP) in the absence of any identifiable cause including intracranial mass, hydrocephalus, or abnormal cerebrospinal fluid (CSF). IIH primarily affects overweight women of childbearing age. Fulminant IIH signifies the severe and rapidly progressive form of IIH.[1,2] Malignant hypertension defined as severe elevations in blood pressure (BP) (>180/120 mm Hg) is a medical emergency and leads to target organ damage.[3] Malignant hypertension causes hypertensive retinopathy, and bilateral optic disc edema is a feature of grade 4 hypertensive retinopathy.

Concurrent fulminant idiopathic intracranial hypertension and malignant arterial hypertension are uncommon.[4] In such cases, the diagnosis of fulminant IIH may be overlooked or delayed given the observed bilateral optic disc edema may be attributed to hypertensive optic neuropathy.

We report 4 patients with severe visual loss from concurrent fulminant IIH and malignant hypertension where the delay in the diagnosis of IIH resulted in poor visual outcomes. The cases underscore the importance of timely consideration of the possibility of fulminant IIH in patients with malignant hypertension and optic disc edema, particularly in those with progressive visual loss. Prompt work-up with brain MRI with contrast and MR venogram to detect venous sinus thrombosis and neuroimaging signs of IIH as well as consideration of MR arteriogram to detect dural arterio-venous fistula followed by a lumber puncture with

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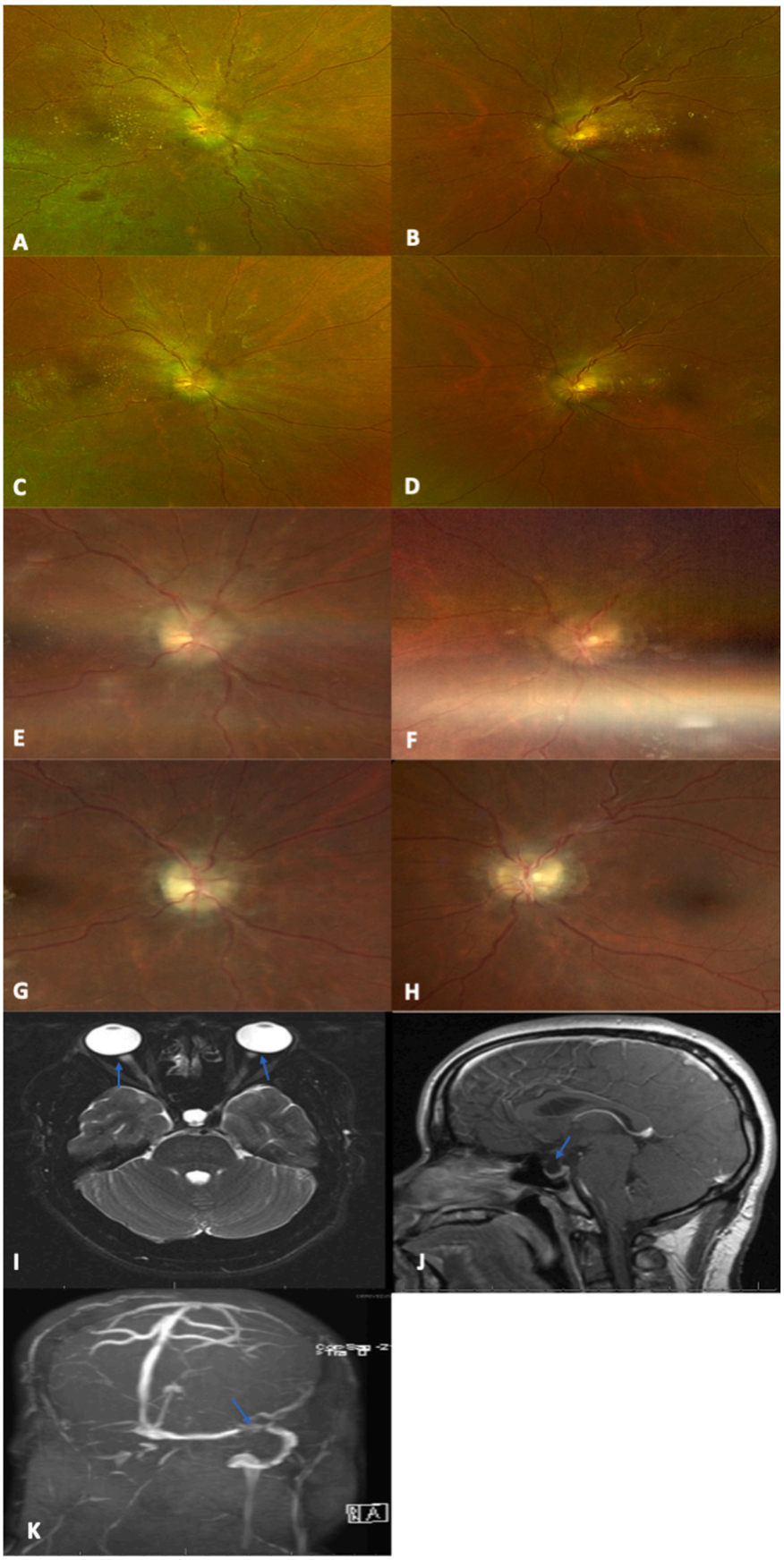

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Fig. 1. Case 1. (A, B) Optic disc edema, macular exudates, and retinal hemorrhages on initial ocular examination. (C, D) Persistent changes 2 months later. (E, F) Persistent changes 4 months after the initial examination. (G. H.) Optic disc pallor, reduced edema, hemorrhages, and exudates after ventriculoperitoneal shunt. months after the onset of vision loss. (I, J, K) MRI and MRV showing mild prominence of cerebrospinal fluid at the optic nerve head, empty sella, and right-sided dominant venous sinus system with focal transverse sinus narrowing and no evidence of thrombosis.

opening pressure are essential to initiate prompt treatment of IIH.

2. Findings

2.1. Case 1

A 31-year-old African American man with a BMI of 25 kg/m^2, uncontrolled hypertension, and chronic kidney disease presented to a local emergency department (ED) with a 2-month history of headache and gradual progressive blurry vision. BP was 250/160, and he was admitted for BP management. After discharge, BP was 156/95, but vision remained blurry. Two weeks later, at our ocular ED, visual acuity (VA) was counting fingers in the right eye and light perception in the left eye. Hypertensive retinopathy and bilateral optic disc edema with nasal macular exudates, and flame-shaped hemorrhages were found and attributed to grade 4 hypertensive retinopathy (Fig. 1A and B). Two months later, VA remained unchanged, optic nerves showed less edema bilaterally with collateral blood vessels on the left, reduced macular exudates, and resolution of flame-shaped hemorrhages (Fig. 1C and D). Two months later, the patient returned to our ocular ED with light perception vision in both eyes and persistent bilateral optic disc edema (Fig. 1E and F). BP was 134/82, and brain MRI with contrast and MR venogram revealed signs of increased intracranial pressure (ICP) (Fig. 1I, J, 1K). Lumbar puncture (LP) opening pressure was 43 cm water with normal cerebrospinal fluid (CSF) analysis. Urgent ventriculoperitoneal shunt was performed and furosemide was initiated given poor renal function precluded the use of acetazolamide. VA improved to counting fingers bilaterally and 3 months later, VA was stable with bilateral pale optic discs (Fig. 1G and H).

2.2. Case 2

A 37-year-old healthy Caucasian man with a BMI of 30 kg/m^2 was evaluated locally for progressive headaches and blurry vision for 6 weeks. BP was 220/180 and bilateral disc edema was found. He was admitted through a local ED for uncontrolled BP and signs of acute kidney dysfunction. One month later, bilateral disc edema and hypertensive retinopathy were documented at a local ophthalmic clinic; BP control and a 2-month follow-up were recommended. However, one month later, he visited our ocular ED due to progressive vision dimming. BP was 115/87, VA was 20/100 in each eye, and bilateral optic disc edema with peripapillary retinal hemorrhages and exudates (Fig. 2A and B) were found with severely constricted visual fields (Fig. 2E and F). Brain MRI and MR venogram revealed signs of increased intracranial pressure (Fig. 2 K, L, M, N). LP opening pressure was 40 cm water with normal CSF analysis. Urgent bilateral optic nerve sheath fenestrations and acetazolamide (1 g twice daily) were initiated given renal dysfunction, and the visual fields improved (Fig. 2G and H). After 1 month, VA worsened to 20/200 bilaterally with worsening visual field (Fig. 2I and J). A ventriculoperitoneal shunt was placed, and acetazolamide was increased to 2 g twice daily. One year later, visual acuity was count-fingers in the right eye and 20/400 in the left eye with pale optic discs bilaterally (Fig. 2C and D), and visual fields remained constricted.

2.3. Case 3

A 30-year-old African American man with a BMI of 35.8 kg/m^2 had intermittent blurry vision and headaches for 3 months. Initially, evaluation at a local ED showed a BP of 214/159. He was admitted for BP management, and no ocular exam was recorded. One month later at our neuro-ophthalmology clinic, the patient reported worsening vision bilaterally. BP was 132/78, and visual acuity was 20/40 in the right eye and 20/100 in the left eye with bilateral optic disc edema, retinal hemorrhages, and macular exudates bilaterally (Fig. 3A and B). Visual fields size 5 showed severe generalized depression bilaterally (Fig. 3C and D). Brain MRI and MR venogram revealed empty sella and transverse venous sinus narrowing consistent with increased ICP. Urgent LP showed an opening pressure of 43 cm water with normal CSF analysis. Acetazolamide 1000 mg twice daily was initiated, and optic nerve sheath fenestration bilaterally were performed. Subsequently, ventriculoperitoneal shunt placement was performed due to progressive visual loss, and acetazolamide was increased to 1000 mg three times daily. Vision stabilized to hand motion bilaterally one month after a ventriculoperitoneal shunt.

2.4. Case 4

A 39-year-old African American woman with a BMI of 19.5 kg/m^2 and a history of heart failure presented to a local ED with tinnitus, blurry vision, and headache. BP was 278/180 and led to admission. An ophthalmic exam was not recorded. After BP control, she was discharged and instructed to follow at an ophthalmic clinic. She missed the follow-up visit and visited our ocular ED four months later. BP was 206/104, and VA was hand motions in the right eye and 20/200 in the left eye. Bilateral optic disc edema with tortuous blood vessels was observed (Fig. 4A and B). The brain MRI and MR venogram revealed empty sella and transverse venous sinus narrowing, consistent with increased ICP. Lumbar puncture indicated an opening pressure of 43 cm water with normal CSF analysis. An MRI of the thoracic spine was performed to rule out spinal and spinal cord abnormalities given her low BMI and showed no significant findings. She underwent bilateral optic nerve sheath fenestrations, and acetazolamide at 1 g three times daily was initiated. Following the treatment, her headache and tinnitus improved, and her vision stabilized with pale discs bilaterally (Fig. 4C and D).

3. Discussion

We report a case series of 4 patients with severe progressive vision loss with concurrent fulminant IIH and malignant hypertension, and the demographics, clinical course, and outcomes are summarized in Table 1. The diagnosis of IIH was delayed given the observed bilateral optic disc edema in all 4 patients was initially attributed to hypertensive vasogenic optic edema from grade 4 malignant hypertension. The average time of delay was 3 months (range 1–4 months) from initial presentation to the diagnosis of IIH which was subsequently confirmed based on MRI, MRV, and LP CSF opening pressure (40–43 cm water) with normal CSF analysis. Over 90 % of IIH patients are women with high BMI,[5] but of the 4 cases, 3 were men and the BMI ranged from 19.5 to 36 kg/m^2. Our case series highlights the need to promptly include IIH in the differential diagnosis in patients with bilateral optic disc edema including patients with malignant hypertension, particularly in those experiencing progressive visual loss, regardless of gender or BMI.

Progressive visual loss in malignant hypertension with disc edema should prompt the clinician to initiate work-up for other causes including intracranial hypertension. The bilateral optic disc edema from malignant hypertension and papilledema may have similar features including flame-shaped hemorrhages, congested retinal veins, and exudates. However, visual function in hypertensive vasogenic optic edema is generally preserved or mildly affected.[6] The pathophysiology of hypertensive vasogenic optic edema is primarily attributed to vascular

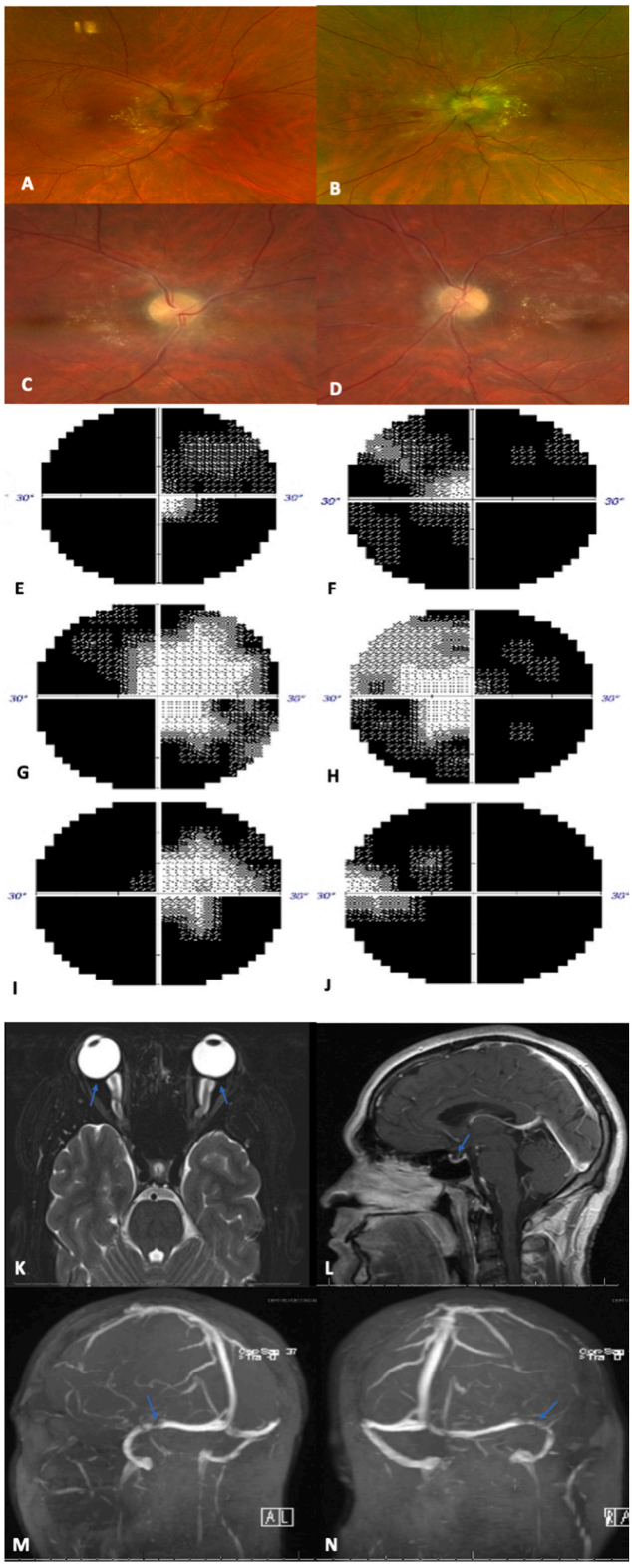

Fig. 2. Case 2. (A, B) Optic disc edema, peripapillary exudates, and flame hemorrhage 3 months after onset of vision loss. (C.D) Optic disc pallor 1 year later after IIH treatment. (E, F) Severely constricted visual fields 3 months after onset of vision loss. (G, H) Improved visual fields after bilateral optic nerve sheath fenestrations and acetazolamide therapy. (I, J) Worsened visual fields 1 month later prompting ventriculoperitoneal shunt. (K, L, M, N) MRI and MRV showed prominent cerebrospinal fluid at the optic nerves, empty sella, bilateral focal transverse sinus narrowing, and no evidence of thrombosis.

leakage and is not caused by increased intracranial pressure.[7] Conversely, malignant hypertension is not a feature of fulminant IIH. Thambisetty and colleagues reported 16 cases of fulminant IIH, with 4 cases showing well-controlled hypertension, and none had malignant hypertension.[2] In our case series, patients 1 and 2 visited ophthalmologists within one month of their initial ED visits due to worsening vision, but the possibility of IIH was not considered. The diagnosis of IIH was only made months later when they visited an ocular ED due to further vision decline.

The lack of prompt eye examinations also contributed to the delay in the diagnosis of fulminant IIH in our patients. Three out of the four patients (Cases 1, 3, 4) did not receive eye exams during their initial ED visits, despite symptoms of vision loss. The ED physicians may have focused on life-threatening conditions associated with malignant hypertension and attributed visual complaints to hypertensive retinopathy. Adequate funduscopic examination is rarely performed by ED physicians, even though nonmydriatic ocular fundus photography has been proven to be effective in identifying and monitoring fundus abnormalities in the ED setting.[8] Prompt identification of severe optic disc edema and urgent brain MRI with contrast and MR venography or CT venogram would have identified signs of increased intracranial pressure. In particular, focal narrowing of the transverse sinus is considered one of the most sensitive neuroradiologic signs of IIH.[9] MR venography or CT venogram is also essential to detect venous sinus thrombosis. MR arteriogram may be needed to detect dural arterio-venous fistula as a secondary cause of elevated intracranial pressure.[10] Although MR arteriogram was not performed in this retrospective case series, there were no abnormal MRI signs to suggest dural arterio-venous fistula, such as parenchymal ischemia, parenchymal edema, or abnormal venous structures.

4. Conclusion

Our case series underscores the importance of considering IIH in the differential diagnosis and prompt work-up in patients with malignant hypertension with bilateral optic edema and progressive visual loss regardless of gender or BMI.

CRediT authorship contribution statement

Bashaer Aldhahwani: Writing – review & editing, Writing – original draft, Methodology, Conceptualization. **Serena M. Shah:** Writing – review & editing, Writing – original draft. **Hong Jiang:** Writing – review & editing, Visualization, Supervision. **Byron L. Lam:** Writing – review & editing, Visualization, Supervision, Methodology, Conceptualization.

Patient consent

Written consent to publish these cases has not been obtained. This report does not contain any personal identifying information.

Conflict of interest

The authors have no financial disclosures: BA, SMS, HJ, or BLL.

Authorship

All authors attest that they meet the current ICMJE criteria for authorship.

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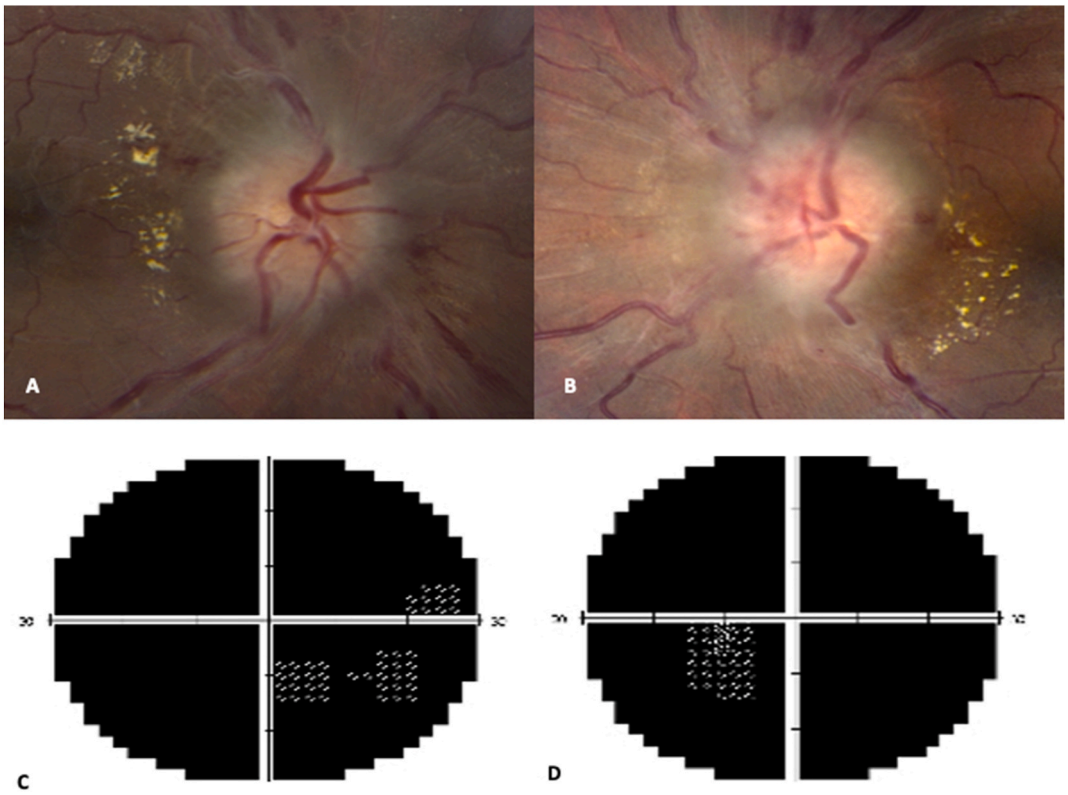

Fig. 3. Case 3. (A, B) Optic disc edema, macular exudates, and retinal hemorrhages on initial ocular examination. (C, D) Visual fields size 5 showed severe generalized depression bilaterally on initial examination.

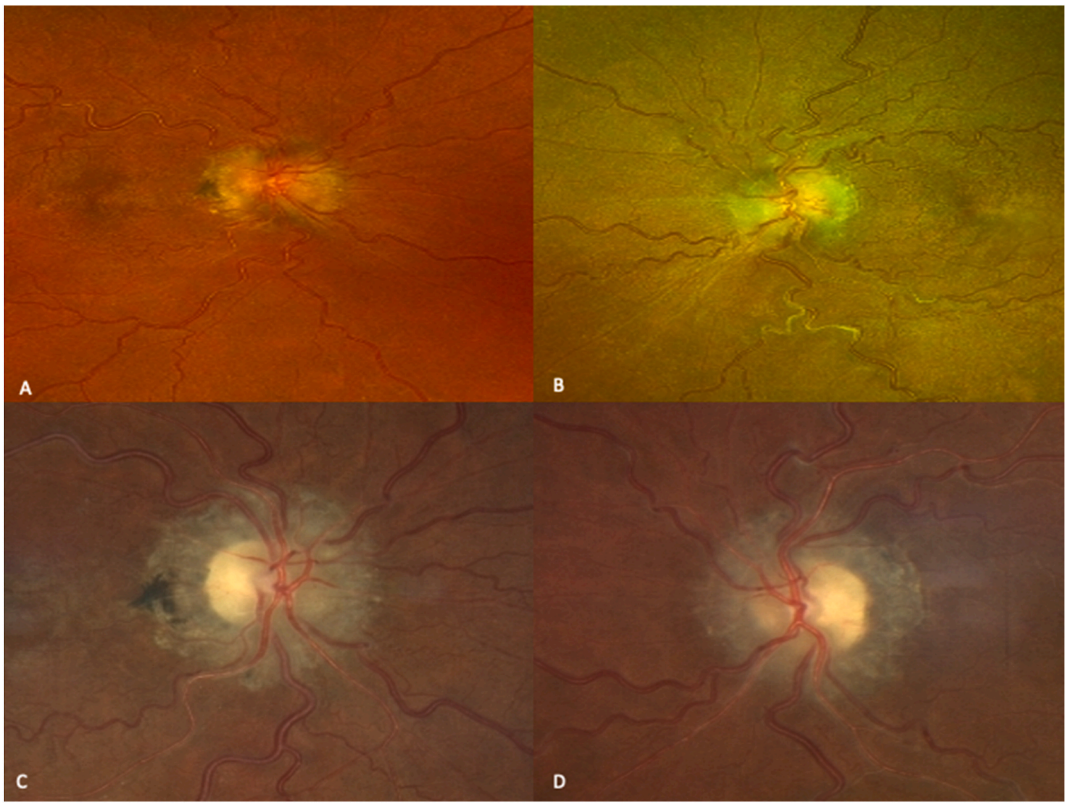

Fig. 4. Case 4. (A, B) Bilateral optic disc edema with tortuous blood vessels on initial ocular examination. (C, D) Optic disc pallor with edema after bilateral optic nerve sheath fenestrations and acetazolamide therapy.

Table 1
Demographics, Clinical course, and Outcomes of 4 Patients with Concurrent Fulminant Idiopathic Intracranial Hypertension and Malignant hypertension.

Case	Age / Sex	BMI	Presenting BP & Comorbidities	Presenting Symptoms	Initial BCVA & Ocular Findings	Time to IIH diagnosis	Brain MRI & MRV	CSF opening pressure	Treatment	Final BCVA
1	31 M	25.0	250/160, chronic kidney disease	Progressive bilateral blurry vision, headaches	LP OU, disc edema & exudates, macular exudates & intra-retinal hemorrhages OU	4 months	Empty sella, focal transverse sinus narrowing	43 cm H_2O	VP Shunt, furosemide	CF OU
2	37 M	30.0	220/180	Bilateral "darkening of vision", headaches	VA 20/100 OU, disc edema & exudates OU, constricted VF OU	3 months	Empty sella, focal transverse sinus narrowing, Prominent optic nerve CSF	40 cm H_2O	ONSF OU VP Shunt, acetazolamide	CF OD, 20/400 OS
3	30 M	35.8	214/156, chronic kidney disease	Headaches	20/40 OD, 20/100 OS, disc edema, peripapillary retinal hemorrhages & exudates, generalized depressed VF	1 month	Empty sella, focal transverse sinus narrowing	43 cm H_2O	ONSF OU VP Shunt, acetazolamide	HM OU
4	39 F	19.5	278/180, heart failure	Bilateral progressive vision loss, headaches, tinnitus	HM OD, 20/200 OS, disc edema with pallor	4 months	Empty sella, focal transverse sinus narrowing	43 cm H_2O	ONSF OU, acetazolamide	HM OD, 20/400

BMI = body mass index; BCVA = best-corrected visual acuity; BP = blood pressure; F = female; IIH = idiopathic intracranial hypertension; M = Male; MRI = magnetic resonance imaging; MRV = magnetic resonance venography; OD = right eye; OS = left eye; OU = both eyes; ONSF = Optic Nerve Sheath Fenestration; VF = Visual Fields; VP = Ventriculoperitoneal; Y = Years.

Declaration of competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

The authors have no conflict of interest.

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