

Idiopathic intracranial hypertension presenting as bilateral palpebral venous engorgement

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ABSTRACT

Purpose: To report the case of a woman in her fifties whose presenting symptom of idiopathic intracranial hypertension was engorgement of the eyelid veins.

Observations: Bilateral engorged palpebral veins were visible through the skin. Dilated fundus examination revealed bilateral optic disc edema. Magnetic resonance imaging revealed bilateral posterior globe flattening and empty sella. Magnetic resonance venogram revealed unilateral transverse sinus stenosis. Lumbar puncture revealed elevated opening pressure. All other initial diagnostic tests were negative.

Conclusions and importance: Idiopathic intracranial hypertension (IIH) typically presents with headache, vision changes, and weight gain. Palpebral venous engorgement has to the authors' knowledge not been previously reported as a presenting sign of IIH. Physicians should recognize this finding and its potential association with elevated intracranial pressure.

1. Introduction

Idiopathic intracranial hypertension (IIH) is a disorder of elevated intracranial pressure of unknown etiology. The typical patient presents between ages 20–50, is female, and has an elevated body mass index (BMI). Patients commonly present with headache, transient visual obscurations, and pulsatile tinnitus.¹ Papilledema is the main ocular finding.¹ Workup must include a broad evaluation for vascular, inflammatory, infectious, and neoplastic disease prior to labelling the condition idiopathic. Ultimately, diagnosis is made with lumbar puncture demonstrating high intracranial pressure (>20 mmHg). Brain imaging can also be of utility, typically revealing the signs of elevated intracranial pressure such as flattened posterior globes and empty sella. Treatment is aimed at lowering intracranial pressure by first removing any inciting agents (i.e., antibiotics or hormones), encouraging weight loss, and utilizing medications such as oral carbonic anhydrase inhibitors. If these methods fail to control disease, procedures can be implemented such as optic nerve sheath fenestration,

ventriculoperitoneal shunt, or venous sinus stenting.^{1–5.}

We share a case of IIH with an atypical presentation. To our knowledge, this is the first report of IIH presenting with engorged palpebral veins.

2. Case report

A female in her fifties presented to her primary care physician (PCP) for an annual examination when the PCP noticed venous prominence in both upper eyelids and referred her to ophthalmology. The patient reported to her ophthalmologist progressive worsening of enlarged vessels around her eyes for six months. When asked about vision changes, she acknowledged occasional spots in her vision when bending over for the past several months, and intermittent blurry vision of the left more than right eye, noted to be worse in the morning. She had occasional dull headache, and when asked about ringing in her ears did admit to occasionally hearing her heartbeat in her head. There was no significant past medical history or medications. Weight was 130 lb., BMI 29.6 (normal 18.5–24.9). Visual acuity, intraocular pressure, extraocular

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Abbreviations

IIH =	Idiopathic intracranial hypertension
OCT =	ocular coherence tomography
RNFL =	retinal nerve fiber layer
MRI =	magnetic resonance imaging
MRV =	magnetic resonance imaging venogram



Fig. 1. Initial Presentation
The patient presented with superior palpebral venous engorgement bilaterally.

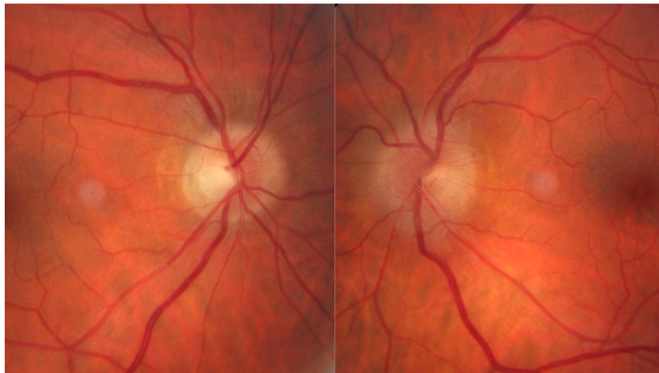


Fig. 2. Optic Disc Edema
Disc photos demonstrating grade 1 optic disc edema in the right eye and grade 2 disc edema in the left eye.

movement, confrontational visual field, and pupil exam were normal. There was right greater than left tortuous veins of the upper eyelid (Fig. 1). Hertel exophthalmometry revealed no frank proptosis (20 mm right eye, 19 mm left eye; normal <21). The anterior segment was normal. Examination of the posterior segment revealed possible mild right optic disc edema and definite left optic disc edema (Fig. 2). Blood pressure was normal. Cranial nerves were intact and there were no focal neurological deficits.

Optical coherence tomography of the retinal nerve fiber layer (RNFL) revealed normal nerve thickness bilaterally (Fig. 3). Humphrey visual field 24-2 demonstrated a few nonspecific scattered defects bilaterally (Fig. 4). Magnetic resonance imaging (MRI) of the brain and orbits with and without intravenous contrast was obtained, revealing bilateral

posterior globe flattening (Fig. 5a), bilateral optic nerve sheath thickening and tortuosity (Fig. 5b), and a partially empty sella. Magnetic resonance venogram (MRV) revealed a left dominant transverse venous sinus with mild stenosis (Fig. 6). Lumbar puncture was performed with elevated opening pressure at 26 cm, normal cell counts and protein, and negative culture. The serum was tested for Lyme disease, angiotensin converting enzyme, lysozyme, and anti-neutrophil cytoplasmic antibodies, all of which were negative.

After thorough consideration, a diagnosis of IIH was made given the absence of an identifiable etiology and the presence of characteristic imaging findings including empty sella, flattened globes, and venous sinus stenosis.

The patient was started on oral acetazolamide with subsequent improvement in disc edema, headache, and transient visual obscurations but with persistence of palpebral venous engorgement. She developed bothersome peripheral neuropathy as a side effect of acetazolamide and wished to stop the medication. She then underwent venous sinus manometry which revealed a positive pressure gradient of 16 mmHg in the stenotic transverse sigmoid junction. A unilateral venous sinus stent was placed at the area of stenosis by interventional neuroradiology. At one and three months postop, the patient reported improvement in her headache but persistence of eyelid venous engorgement.

3. Discussion

This unusual case describes a woman referred for ophthalmic evaluation of bilateral eyelid vein engorgement. Her symptoms were associated with dull headache and transient visual obscurations. Dilated fundus examination revealed optic disc edema. MRI revealed flattened globe and empty sella. MRV revealed unilateral transverse venous sinus stenosis. Lumbar puncture revealed elevated opening pressure. The Modified Dandy Criteria⁶ were met, and the patient was diagnosed with IIH.

There have been no reports to our knowledge of IIH presenting as eyelid changes. A review of the anatomy explains how cerebral venous congestion (seen in IIH) impair venous return and lead to upstream venous congestion manifesting in the eyelids. The superior palpebral vein drains into the supraorbital and supratrochlear veins which drain into the superior orbital vein and ultimately into the cavernous sinus (Fig. 7). Pressure elevation in these venous pathways could cause upstream venous congestion and thus eyelid venous engorgement. One might expect to see superior orbital vein dilation in such a setting, a finding which was not observed in our patient. It is reasonable to speculate that perhaps the chronicity of congestion (6 months) allows for a normal radiographical appearing superior orbital vein. Additionally, engorgement of the facial veins to compensate for the congestion, as seen in this patient, may allow for a more normal sized superior orbital vein.

The patient in this report was found to have unilateral transverse venous sinus stenosis. There is an association between intracranial pressure and venous sinus stenosis. Physiologically, this makes sense. The dural venous sinuses collect both venous blood and cerebrospinal fluid (CSF), channeling volume back to the heart. Sinus narrowing impairs proximal venous outflow and CSF resorption, thus contributing to elevated intravenous and intracranial pressure. A positive feedback loop is created, as the elevated intracranial pressure contributes to further elevating the pressure within the stenotic sinus.⁷

Over the past decade there has been substantial literature demonstrating an association between venous sinus stenosis and IIH and

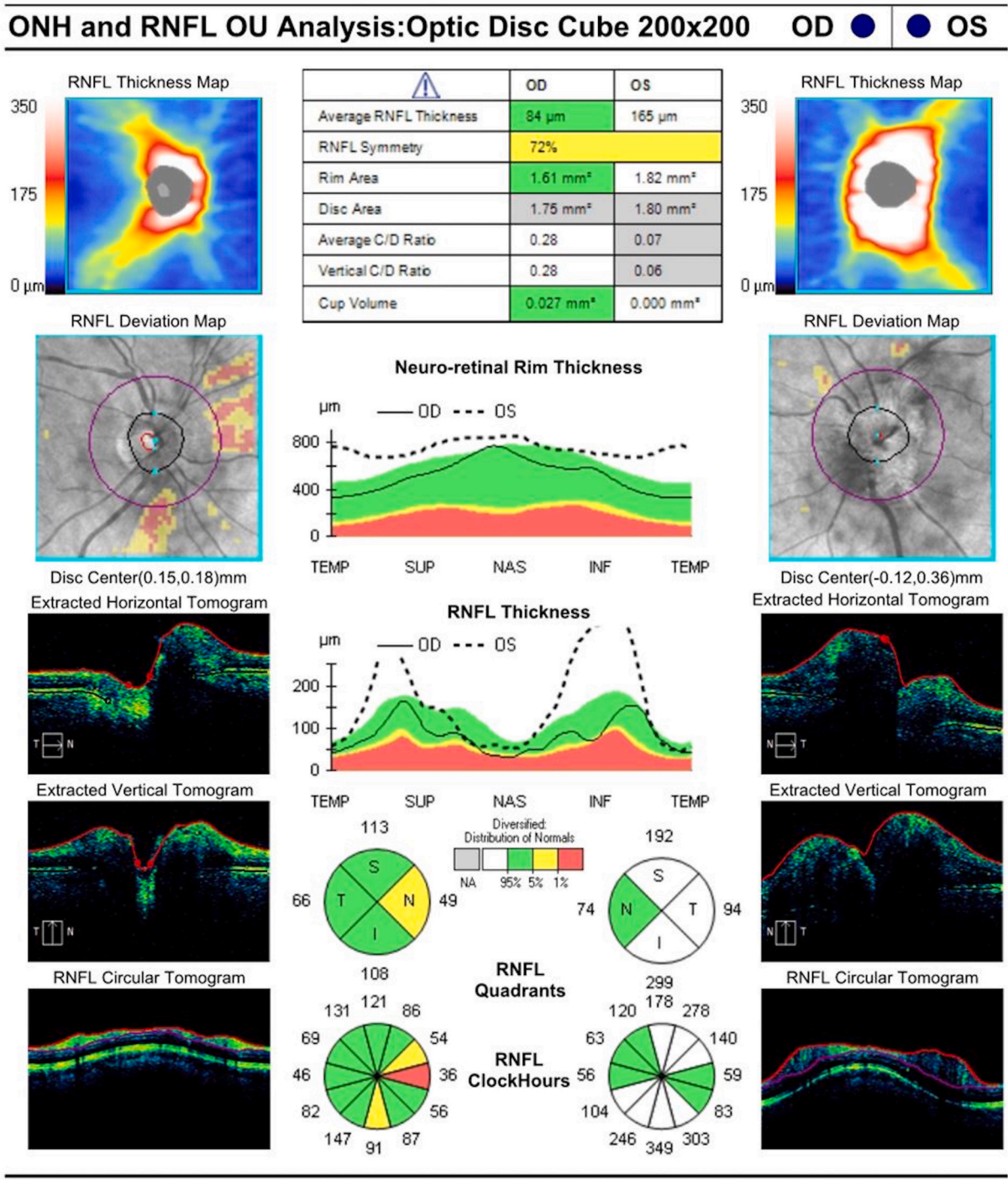


Fig. 3. Retinal Nerve Fiber Layer
Optical Coherence Tomography: Retinal Nerve Fiber Layer is within normal limits on the right but thickened on the left.

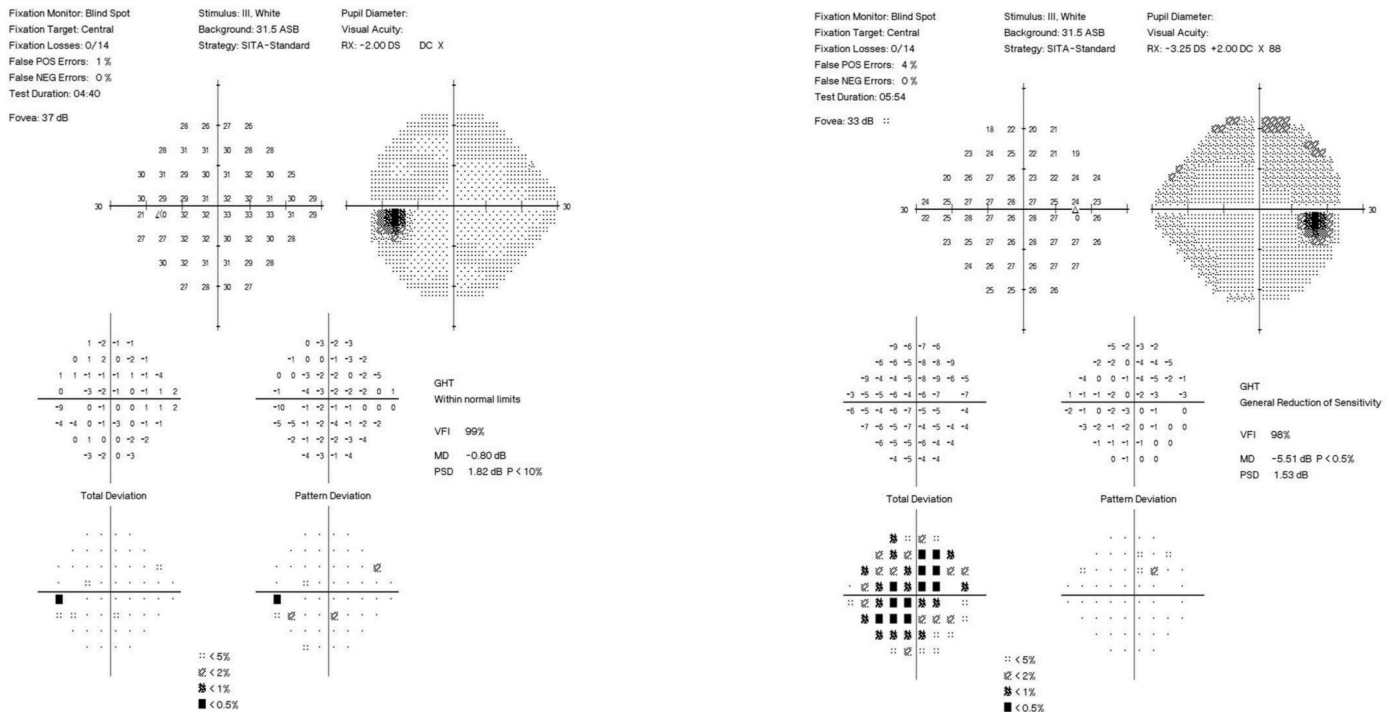


Fig. 4. Humphrey Visual Field
Humphrey Visual Field 24-4 reveals non-specific scattered defects bilaterally.

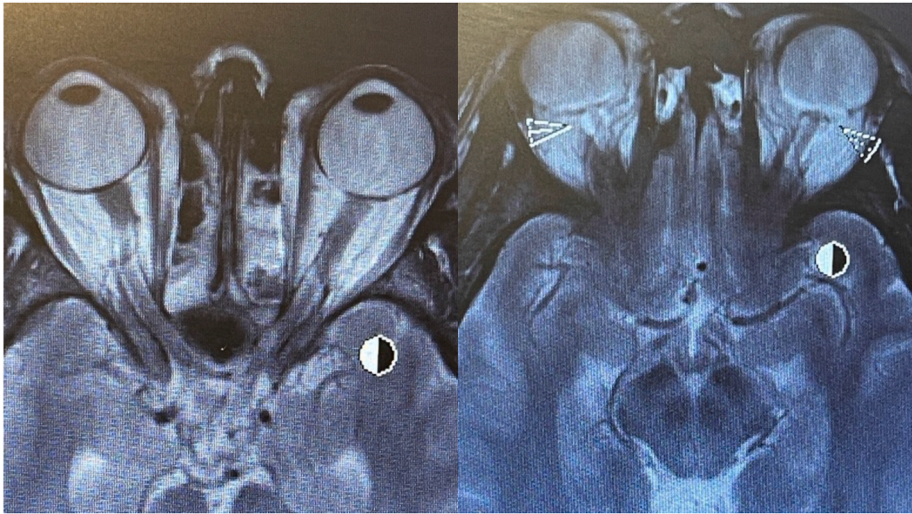


Fig. 5. Magnetic Resonance Imaging
An axial cut of the Magnetic Resonance Imaging orbits with contrast reveals flattened posterior globes (left image) and bilateral optic nerve sheath thickening and tortuosity (right image, arrow heads), classically associated with elevated intracranial pressure.

demonstrating efficacy of sinus stenting for the treatment of IIH.⁷⁻¹¹ Although these studies are potentially biased, as authors are frequently also practitioners of venous sinus stenting, they offer compelling data. Published in the American Journal of Neuroradiology, Morris et al. reported transverse venous sinus stenosis in 94 % of IIH patients, while this finding was present in 3 % of controls.⁸ Stenting is considered when

1) IIH is refractory to medical management, 2) venous sinus stenosis is present with a gradient of 8 mmHg or more across the stenotic area, 3) there is no contraindication to dual antiplatelet therapy, and 4) the patient is symptomatic.⁷
The patient in this report met the criteria for stenting and underwent uncomplicated stent placement in her stenotic left transverse venous



Fig. 6. Magnetic Resonance Venogram
Magnetic resonance venogram shows a left dominant transverse venous sinus with stenosis (black arrow)".

sinus. At her three month follow up, she reported resolution of headaches and pulsatile tinnitus, demonstrated improvement in optic disc edema, but showed no changes in the eyelid venous engorgement.

4. Conclusions

IIH typically presents with headache, vision changes, and weight gain. There have been no previously documented reports of IIH presenting with engorgement of the palpebral veins. Ophthalmologists should be aware of this unusual presentation of IIH.

Patient consent

Written consent to publish personal information, case details, and photos of the face that may be identifiable was obtained from the patient.

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Authorship

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

CRediT authorship contribution statement

Samantha D. Butterfield: Writing – original draft, Visualization, Methodology, Investigation, Funding acquisition, Data curation,

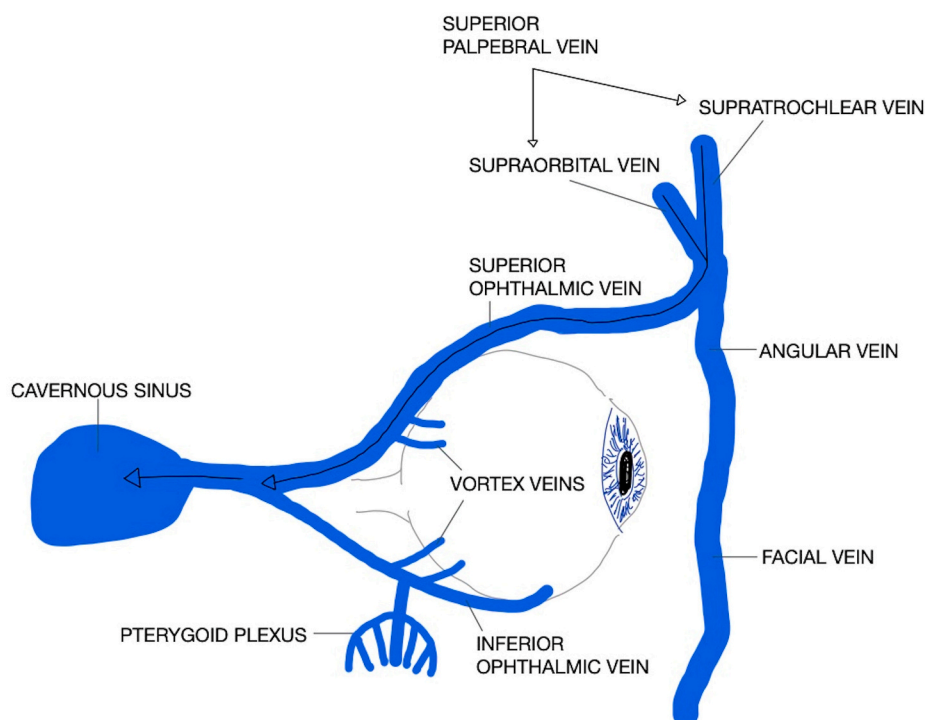


Fig. 7. Venous drainage of the eyelid and orbit.
Adapted from Netter, Atlas of Human Anatomy, 5th edition, Saunders, 2011, ISBN 978-1-4160-5951-6, Plate 85.

Conceptualization. **Rona Z. Silkiss:** Writing – review & editing, Supervision, Resources, Methodology, Investigation, Conceptualization.

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.

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