

A Unique Case Report on “Bilateral Optic Neuropathy as a Rare Manifestation of Spontaneous Intracranial Hypotension”

K SANKAVI GAUTHAM*, CJ SELVAKUMAR†, V SADEESH KUMAR‡, M SACRATIS§, N SHOBANA#

ABSTRACT

Spontaneous intracranial hypotension (SIH) is an uncommon condition characterized by low cerebrospinal fluid (CSF) volume through a dural defect leakage, which results in multiple debilitating clinical manifestations. Although ocular manifestations are common, vision impairment due to SIH is not commonly reported. Optic neuropathy is a rare yet significant complication of SIH. This case documents the association between optic neuropathy and SIH, highlighting the diagnostic challenges and the complexities of managing this condition.

Keywords: Orthostatic headache, bilateral optic neuropathy, subdural hygroma, dural venous thrombosis, epidural blood patch

The clinical characteristics of spontaneous intracranial hypotension (SIH) often result from stretching of the intracranial and cervical dura and dilatation of intracranial venous structures and dural sinuses resulting in orthostatic headache, neck pain, and radicular symptoms¹. The common ocular manifestations include blurry vision, double vision, and ophthalmoparesis². Visual field deficits are related to stretching of optic nerve over pituitary fossa or vascular congestions of the optic nerve. Optic neuropathy in SIH is a rare and underreported phenomenon. Previously one case reported an association between monocular optic neuropathy and SIH¹. This is the first case documenting the association of bilateral optic neuropathy in SIH and sheds light on this infrequent complication.

CASE REPORT

A 21-year-old female with no known comorbidities presented with the complaints of insidious-onset,

gradually progressive orthostatic headache for 1 year. The headache was holocranial, throbbing type of moderate to severe intensity associated with neck pain. Headache had gradually evolved into nonorthostatic chronic, persistent headache for 5 months. The headache was triggered by coughing and abrupt head movements. She also noticed gradually progressive painless diminution of vision of both eyes (right more than left [R > L]) in the last 4 months with no history of diplopia/lower cranial nerve involvement. There was no relevant medical history or previous trauma or lumbar puncture. On examination, patient was moderately built and nourished. There were no cutaneous or articular stigmata for connective tissue disorder. Cranial nerve examination revealed right eye relative afferent pupil defect (RAPD) (Fig. 1) with perception of light in right eye and visual acuity of 1/60 in left eye. Fundus examination revealed temporal pallor of optic disc (R > L) with well-defined margins with normal retinal vessels with dull macular foveal reflex suggestive of both eye primary optic neuropathy (R > L) (Fig. 2). Ocular movements were full; examination of lower cranial nerves, sensorimotor, cerebellum, and higher mental functions were unremarkable. On investigating, complete blood picture revealed microcytic hypochromic anemia; renal function tests, liver function tests, thyroid profile, urine routine were within limits. Voluntary counseling and testing for human immunodeficiency virus (HIV) was negative. Antinuclear antibody test,

*Resident

†Assistant Professor

‡Associate Professor

#Professor

Coimbatore Medical College Hospital, Coimbatore, Tamil Nadu, India

Address for correspondence

Dr K Sankavi Gautham

009 Manchester Perks II, Uppilpalayam, Coimbatore, Tamil Nadu, India

E-mail: sangavikaarthic@gmail.com

serum homocysteine, antiphospholipid antibody, serum angiotensin-converting enzyme were negative.

Optical coherence tomography revealed thinning of retinal nerve fiber layer of all quadrants in right eye, and superotemporal and inferotemporal quadrants in the left eye. A visual evoked potential (VEP) revealed no response in the right eye and reduced amplitude and prolonged P100 latency in left eye. A lumbar puncture was performed and the cerebrospinal fluid (CSF) opening pressure measured at the lateral decubitus position was 5 cm of H₂O with normal biochemistry and microbiological analysis including cartridge-based nucleic acid amplification test (CBNAAT) and adenosine deaminase (ADA). Noncontrast CT brain revealed thickened, hyperdense dura along the tentorium (Fig. 3). The contrast-enhanced magnetic resonance imaging (MRI) brain with magnetic resonance venography (MRV) revealed engorged dural spaces, enhancement of pachymeninges, flattening of

pons, sagging of bilateral tentorial and cerebellar tonsils (Fig. 4 a-c) with thinning of both optic nerves complicated by chronic dural venous sinus thrombosis.

With the background of orthostatic headache, low CSF opening pressure, neuroimaging suggestive of engorged dural venous sinus and sagging of brain, the probability of SIH was considered. So, a MRI myelogram was done, which demonstrated CSF leak at left lower dorsal level (D11-D12) (Fig. 5). A final diagnosis of SIH was made. Patient was treated with adequate fluids, analgesics, anticoagulants, and subsequently 20 mL epidural blood patch in the lower thoracic region. The headache and general well-being improved and there was no further diminution of visual acuity. Repeat neuroimaging, VEP, visual field charting was planned for during follow-up.

On follow-up, visual field charting showed improvement of visual acuity 1/60 in right eye and 2/60 in left eye. VEP showed P100 latency of 136 ms in right eye and left eye P100 latency of 120 ms with reduced amplitude in both



Figure 1. Right eye RAPD.

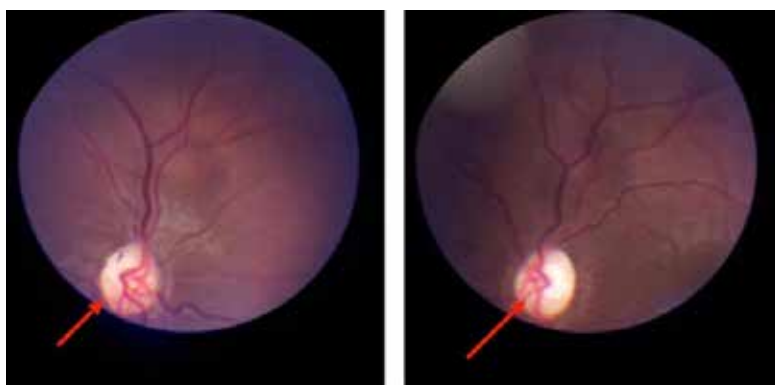


Figure 2. Both eye fundus - pale disc with well-defined margins, normal retinal vessels suggestive of primary optic neuropathy.

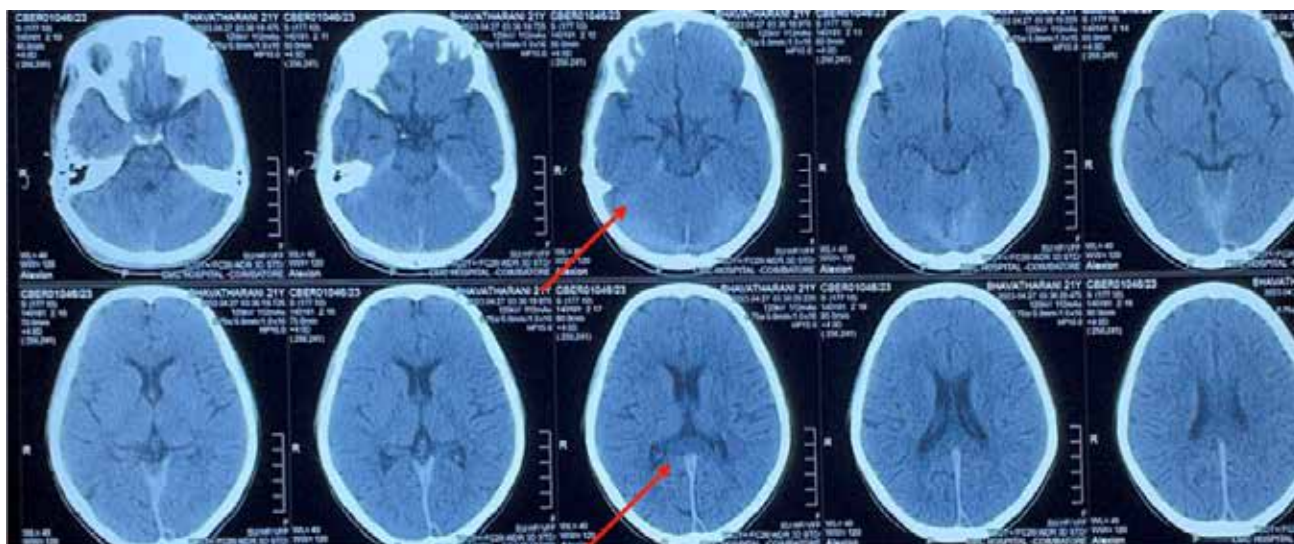


Figure 3. Noncontrast CT brain showing thickened hyperdense dural along posterior falx and tentorium.

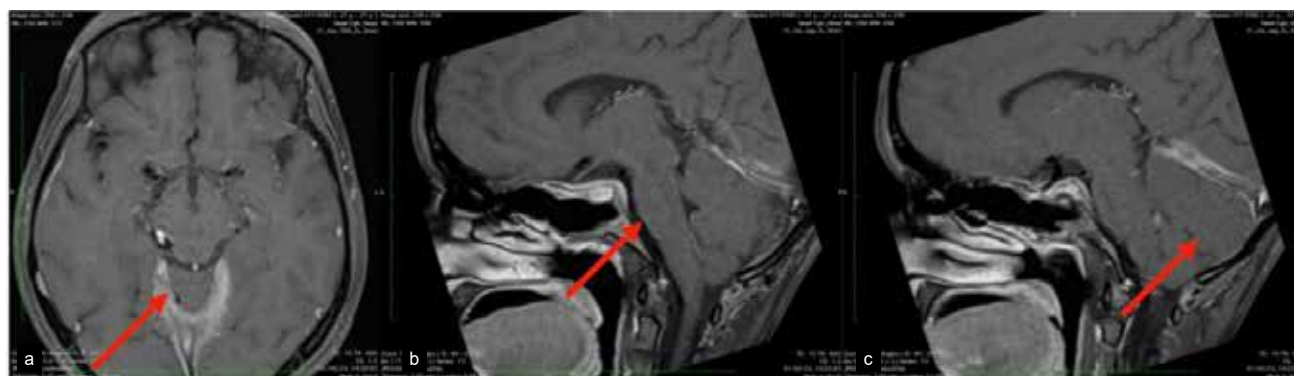


Figure 4. MRI contrast axial-engorged dural venous sinus (a); MRI sagittal contrast flattening of pons (b); and MRI sagittal contrast herniation of cerebellar tonsils (c).



Figure 5. MRI myelography shows CSF leak at lower dorsal level (D11-D12).

eyes. However, MRI showed persistence of sagging of brain and it was planned to repeat in serial follow-ups.

On follow-up visual acuity improved in both eyes. Right eye visual acuity - 20/200 and left eye visual acuity 20/100. Both eye P100 latency is within limits. Neuroimaging findings are static at present.

DISCUSSION

Spontaneous intracranial hypotension is estimated to affect 5 per 1,00,000 people per year with a female preponderance³. It occurs when mechanical stressors (spiculated osteophytes, herniated discs, nerve root diverticula) incite small dural tears causing CSF to leak into extradural space^{4,5}. It can also occur following lumbar puncture, head trauma, spinal shunts, or rarely

spontaneously. It may be exacerbated by connective tissue disorder.

According to Monro-Kellie doctrine, skull is a rigid compartment, which contains brain parenchyma, blood, and CSF compartment¹. CSF exerts a buoyant force suspending the cranial nerves and brain parenchyma and prevents it from sagging downward². These components are balanced in a state of dynamic equilibrium in normal circumstances. When the volume of CSF decreases in SIH, it is compensated by increase in the volume of other components to maintain the equilibrium, which results in venous sinus engorgement, subdural effusion, enlargement of pituitary gland, pachymeningeal enhancement, vascular congestion of optic nerve². This results in traction of intracranial dura resulting in orthostatic headaches, neck pain, and ophthalmoparesis. Similarly, downward traction of optic nerve causing damage to the sheath of the optic nerve along with vascular congestion of the intracranial portion of the optic nerve can account for optic neuropathy. A case of monocular optic neuropathy associated with SIH has earlier been reported, which was attributed to traction/compression and/or vascular congestion of the intracranial portion of optic nerve¹.

This is the first reported case of bilateral primary optic neuropathy in a patient with SIH and this could be attributed to downward traction of optic nerve resulting in optic nerve damage that would eventually result in optic neuropathy.

CONCLUSION

Postural headache and optic neuropathy could be the presenting symptoms of SIH and early recognition is of paramount importance for preserving vision. Our case underscores the importance of early recognition, and prompt intervention in recognizing optic neuropathy in patients with SIH so that we can prevent the potential

consequences of visual impairment or blindness. This also emphasizes the need for the treating physician to be vigilant in dealing with primary optic atrophy and SIH should be considered as one of the differential diagnoses in cases with the relevant clinical background.

Acknowledgments

We would like to extend our sincere gratitude to Professor and Head Dr N Shobana.

REFERENCES

1. Goksel BK, Yildirim T, Sizmaz S, Reyhan M, Karatas M. Optic neuropathy associated with spontaneous intracranial hypotension. *Acta Neurol Belg*. 2012;112(4):361-5.
2. Zada G, Solomon TC, Giannotta SL. A review of ocular manifestations in intracranial hypotension. *Neurosurg Focus*. 2007;23(5):E8.
3. Bond KM, Benson JC, Cutsforth-Gregory JK, Kim DK, Diehn FE, Carr CM. Spontaneous intracranial hypotension: atypical radiologic appearances, imaging mimickers, and clinical look-alikes. *AJNR Am J Neuroradiol*. 2020;41(8):1339-47.
4. Schievink WI. Spontaneous spinal cerebrospinal fluid leaks and intracranial hypotension. *JAMA*. 2006;295(19):2286-96.
5. Hoffmann J. Headache attributed to intracranial hypertension and hypotension. In: Mitsikostas D, Paemeleire K (Eds.). *Pharmacological Management of Headaches*. Headache. Springer, Cham; 2016. https://doi.org/10.1007/978-3-319-19911-5_18

■ ■ ■ ■