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Changes of Peripapillary Hyperreflective Ovoid Mass-Like Structures in a Patient with Idiopathic Intracranial Hypertension

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Abstract **Case Report**

We present a case of idiopathic intracranial hypertension (IIH) in a 30-year-old woman. The ophthalmoscopic examination revealed marked papilledema in both eyes. Magnetic resonance imaging demonstrated bilateral enlargement of the optic nerve sheath and flattened posterior sclera. Optical coherence tomography demonstrated the presence of peripapillary hyperreflective ovoid mass–like structures (PHOMS). On lumbar puncture, the cerebrospinal fluid pressure was 300 mm H_2O. She was prescribed acetazolamide, and the bilateral papilledema gradually improved. Two months later, the papilledema was completely resolved and PHOMS regressed. This case highlights the importance for clinicians to be aware of PHOMS with IIH.

Keywords: idiopathic intracranial hypertension, papilledema, optical coherence tomography, peripapillary hyperreflective ovoid mass–like structures.

INTRODUCTION

Idiopathic intracranial hypertension (IIH) is characterized by increased cerebrospinal fluid (CSF) pressure and papilledema without focal neurologic symptoms [1]. It is a diagnosis of exclusion, and radiologic examinations are traditionally performed to help exclude other causes of intracranial hypertension, such as obstructive hydrocephalus, tumor, chronic meningitis, arteriovenous fistula, internal jugular vein stenosis, and dural sinus thrombosis [1]. The revised diagnostic criteria for IIH were established as follows [2]: (A) papilledema; (B) normal neurologic examination, except for cranial nerve abnormalities; (C) normal brain parenchyma on neuroimaging without evidence of hydrocephalus, mass, or structural lesion, and no abnormal meningeal enhancement on plain and gadolinium-enhanced magnetic resonance imaging (MRI) in typical patients (female and obese), and on magnetic resonance venography for others; if MRI is unavailable or contraindicated, then contrast-enhanced CT may be used; (D) normal CSF composition; and (E) elevated lumbar puncture opening pressure (≥250 mm CSF in adults and ≥280 mm CSF in children [250 mm CSF in awake and normal weight children]) in a correctly performed lumbar puncture. A diagnosis of IIH is definite if the patient fulfills criteria A–E. The diagnosis is considered probable if criteria A–D are met but the CSF pressure is lower than specified for a definite diagnosis.

Peripapillary hyperreflective ovoid mass–like structures (PHOMS) can be observed using optical coherence tomography (OCT) of the optic nerve head [3, 4]. The defining morphologic features of PHOMS include (1) a peripapillary location, abutting on the retina, (2) hyperreflectivity on OCT, (3) an ovoid shape on linear OCT scans through the center of the optic disc, and (4) a mass-like, space-filling structural characteristic of displacing the adjacent retina from the disc [3, 4]. As PHOMS was initially described in the setting of optic nerve head drusen, however, it will be recognized by the association with papilledema/pseudopapilledema, tilted disc syndrome, inflammatory demyelinating disorders and other miscellaneous retinal and optic nerve diseases [3-7]. Herein, we report a case of PHOMS in a patient with IIH.

CASE REPORT

A 30-year-old woman was referred to Jichi Medical University Hospital because of headache. In both eyes, her visual acuity was 1.2, and the anterior segments and ocular pressures were normal. Ophthalmoscopic examination revealed marked papilledema in both eyes **(Fig 1A, B).**

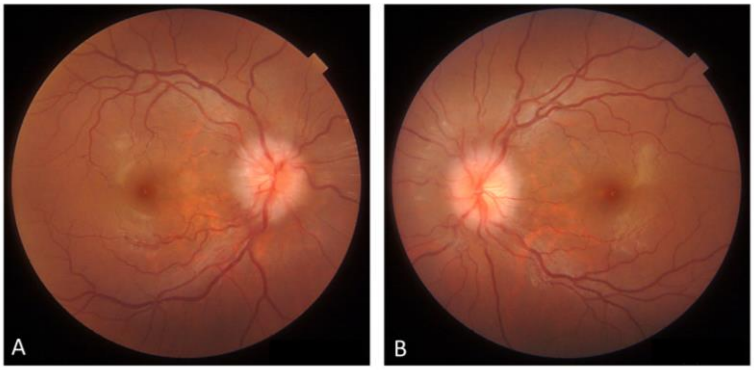

Fig 1: Photographs of the right (A) and left (B) fundus on initial examination

Note: papilledema in both eyes.

The coronal T2-weighted MRI showed bilateral widening of the optic nerve sheaths **(Fig 2A).** The axial T2-weighted MRI revealed tortuosity in the optic nerve and flattened posterior sclera **(Fig 2B).**

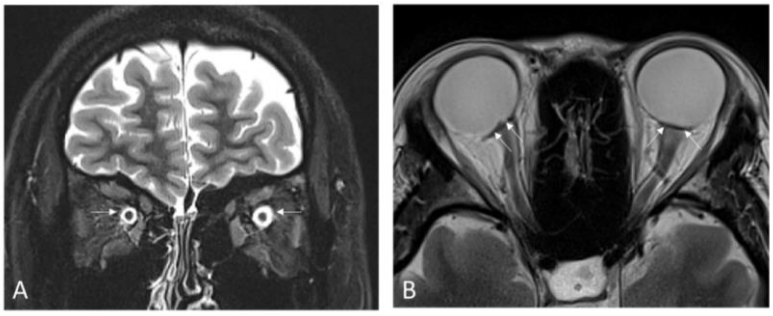

Fig 2: Initial coronal (A) and axial (B) T2-weighted MRI

Note: widening of the optic nerve sheath (A; arrows) and flattened the posterior sclera (B; arrows).

Peripapillary OCT demonstrated PHOMS in both eyes **(Fig 3A, B, arrows).**

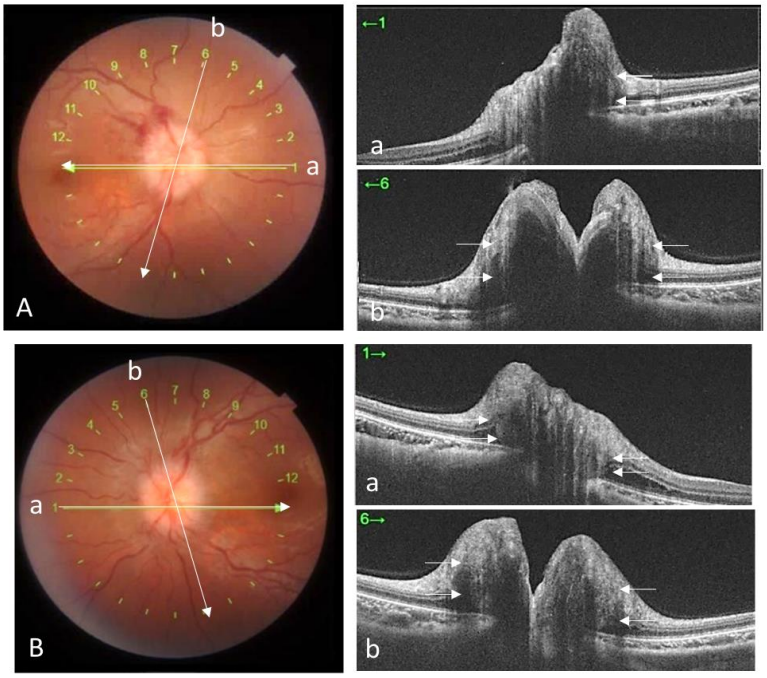

Fig 3: Optical coherence tomography of the right (A) and left (B) eyes

Note: peripapillary hyperreflective ovoid mass-like structures in both eyes (white arrows).

Based on these collective findings, the patient was diagnosed with IIH. A lumber puncture was performed and showed a CSF pressure of 300 mm H_2O and normal CSF composition. The diagnosis of IIH was confirmed, and the patient was prescribed oral acetazolamide 250 mg three times daily. After beginning acetazolamide treatment, the bilateral papilledema gradually improved **(Fig 4).** Two months later, the papilledema was completely resolved (Fig 4), and OCT demonstrated regress of PHOMS **(Fig 5A, B).**

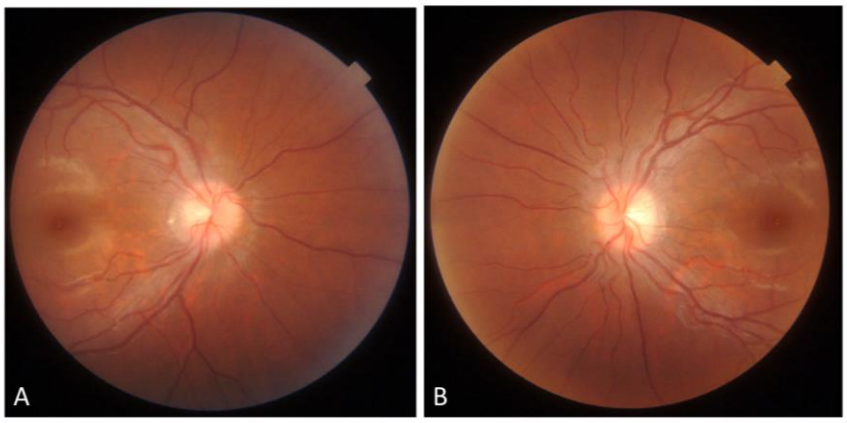

Fig 4: Photographs of the right (A) and left (B) fundus after 2 months of acetazolamide treatment

Note: improvement of papilledema in both eyes.

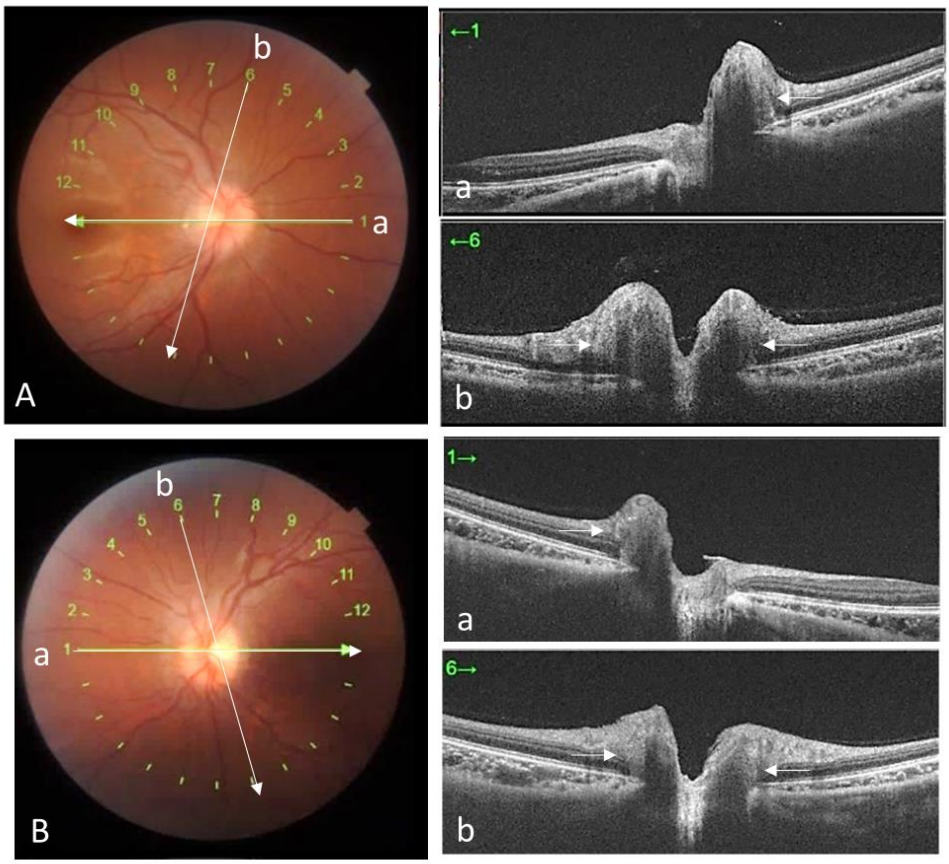

Fig 5: Optical coherence tomography of the right (A) and left (B) eyes after 2 months of acetazolamide treatment

Note: regress of peripapillary hyperreflective ovoid mass-like structures in both eyes (white arrows).

DISCUSSION

The most commonly reported macroscopic findings on MRI of patients diagnosed with papilledema are as follows: (1) enlarged optic nerve sheath, (2) flattened posterior sclera, (3) optic papilla protrusion into the globe, and (4) torturous optic nerve [8-11]. The

present case fulfilled the diagnostic criteria of IIH [2] and showed these MRI findings.

Current theories suggest that IIH is the result of abnormal CSF hydrodynamics; therefore, treatment generally aims to modify CSF production and flow [12]. Medical therapy for IIH typically includes oral acetazolamide, which is a carbonic anhydrase inhibitor with diuretic properties that decreases the CSF [12, 13]. Acetazolamide treatment was successful in this case.

PHOMS is widely believed to be an indicator of impaired axoplasmic flow, which can result from various factors, including mechanical traction from optic disc drusen, papilledema, optic neuritis, and optic disc anomalies, among others [1, 2]. Firstly, on pathological sections of optic disc drusen, degeneration of nerve fiber cells, intracellular and extracellular calcium deposits, and mechanical compression by dense, rock-like calcification can be observed, and axoplasmic flow stasis of nerve fiber axons also occurs [8]. Secondly, studies in primate models of papilledema have shown that the lamina cribrosa's rigid structure compresses the edematous nerve, leading to axoplasmic stasis, as demonstrated using radioactive isotopes [9].

Differentiating pseudopapilledema from optic disc edema caused by intracranial hypertension or other optic neuropathies can be a diagnostic challenge. Pseudopapilledema is a group of optic disc anomalies whose common element is the elevation of the optic disc without true swelling of the axonal fibers. These anomalies include dysplastic optic disc, hypoplastic optic disc, tilted disc, persistent hyaloid remnants, gliosis, myelinated nerve fibers, hamartoma, and drusen [7]. PHOMS can be one of the causes of pseudopapilledema, but the presence of PHOMS does not rule out true papilledema and does not prove pseudopapilledema. Even if the presence of PHOMS is evident, an MRI scan is mandatory to rule out papilledema.

Wibroe *et al*., [14] reported the prevalence of PHOMS in IIH patients. According to their report, PHOMS were found in 81.3% of all IIH patients at least three months after the time of diagnosis. The high prevalence of PHOMS in IIH patients suggest these structures be a result of crowding in the optic nerve head caused by papilledema. In addition, when pressure is normalized and axoplasmatic flow is restored, PHOMS regress in some patients.

CONCLUSION

This case highlights the importance for clinicians to be aware of PHOMS with IIH and we consider PHOMS has potential as non-invasive biomarkers of intracranial pressure.

Disclosure: The authors declare that they have no conflicts of interest in the preparation of this report.

REFERENCES

1. Suzuki, H., Takanashi, J. I., Kobayashi, K., Nagasawa, K., Tashima, K., & Kohno, Y. (2001). MR imaging of idiopathic intracranial hypertension. *American Journal of Neuroradiology*, *22*(1), 196-199.
2. Friedman, D. I., Liu, G. T., & Digre, K. B. (2013). Revised diagnostic criteria for the pseudotumor cerebri syndrome in adults and children. *Neurology*, *81*(13), 1159-1165.
3. Heath Jeffery, R. C., & Chen, F. K. (2023). Peripapillary hyperreflective ovoid mass-like structures: multimodal imaging—A review. *Clinical & Experimental Ophthalmology*, *51*(1), 67-80.
4. Li, B., Li, H., Huang, Q., & Zheng, Y. (2023). Peripapillary hyper-reflective ovoid mass-like structures (PHOMS): clinical significance, associations, and prognostic implications in ophthalmic conditions. *Frontiers in Neurology*, *14*, 1190279.
5. Behrens, C. M., Malmqvist, L., Jørgensen, M., Sibony, P. A., Munch, I. C., Skovgaard, A. M., ... & Hamann, S. (2023). Peripapillary Hyperreflective Ovoid Mass–like Structures (PHOMS) in Children: The Copenhagen Child Cohort 2000 Eye Study. *American Journal of Ophthalmology*, *245*, 212-221.
6. Pratt, L., Rehan, S., West, J., & Watts, P. (2023). Prevalence of peripapillary hyperreflective ovoid mass-like structures (PHOMS) in suspected papilloedema in children. *Eye*, *37*(15), 3209-3212.
7. Mezad-Koursh, D., Klein, A., Rosenblatt, A., Teper Roth, S., Neudorfer, M., Loewenstein, A., ... & Zur, D. (2021). Peripapillary hyperreflective ovoid mass-like structures—a novel entity as frequent cause of pseudopapilloedema in children. *Eye*, *35*(4), 1228-1234.
8. Passi, N., Degnan, A. J., & Levy, L. M. (2013). MR imaging of papilledema and visual pathways: effects of increased intracranial pressure and pathophysiologic mechanisms. *American Journal of Neuroradiology*, *34*(5), 919-924.
9. Brodsky, M. C., & Vaphiades, M. (1998). Magnetic resonance imaging in pseudotumor cerebri. *Ophthalmology*, *105*(9), 1686-1693.
10. Gass, A., Barker, G. J., Riordan-Eva, P., MacManus, D., Sanders, M., Tofts, P. S., ... & Miller, D. H. (1996). MRI of the optic nerve in benign intracranial hypertension. *Neuroradiology*, *38*, 769-773.
11. Jinkins, J. R., Athale, S., Xiong, L., Yuh, W. T., Rothman, M. I., & Nguyen, P. T. (1996). MR of optic papilla protrusion in patients with high intracranial pressure. *American Journal of Neuroradiology*, *17*(4), 665-668.
12. Galgano, M. A., & Deshaies, E. M. (2013). An

update on the management of pseudotumor cerebri. *Clinical neurology and neurosurgery*, *115*(3), 252-259.

13. Demeritt, M., Shechtman, D., & Reynolds, S. (2014). Idiopathic Intracranial Hypertension (IIH): Review &Updates. *Austin J Clin Ophthalmol*, *1*(7), 1033.
14. Wibroe, E. A., Malmqvist, L., & Hamann, S. (2021). OCT based interpretation of the optic nerve head anatomy and prevalence of optic disc drusen in patients with idiopathic intracranial hypertension (IIH). *Life*, *11*(6), 584.