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Peripapillary Hyperreflective Ovoid Mass-Like Structures as a Finding of Pseudopapilledema in Myopic Children with Tilted Disc Syndrome

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Abstract **Case Report**

We describe two cases with peripapillary hyperreflective ovoid mass-like structures (PHOMS) as a finding of pseudopapilledema in myopic children with tilted disc syndrome. ***Case 1***: An 11-year-old boy presenting with suspected papilledema in left eye. The patient had moderate myopia in both eyes. Ophthalmoscopic examination revealed optic disc edema and tilted disc in left eye. Peripapillary optical coherence tomography (OCT) demonstrated PHOMS in left eye that was located in the nasal sector. ***Case 2***: A 15-year-old boy presenting with suspected papilledema in left eye. The patient had mild myopia in both eyes. Ophthalmoscopic examination revealed optic disc edema and tilted disc in left eye. Peripapillary OCT demonstrated PHOMS in left eye that was located in the nasal sector. Clinicians should be aware of PHOMS when a patient presents with myopic eyes and tilted disc syndrome, as in these cases.

Keywords: Papilledema, peripapillary hyperreflective ovoid mass-like structures (PHOMS), optical coherence tomography (OCT), myopia, tilted disc syndrome.

INTRODUCTION

Peripapillary hyperreflective ovoid mass-like structures (PHOMS) can be observed using optical coherence tomography (OCT) of the optic nerve head [1-5]. 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 [1-5]. These PHOMS can cause pseudopapilledema. Here, we present two cases with suspected papilledema.

CASE PRESENTATION

Case 1:

An 11-year-old boy presenting with suspected papilledema in left eye was referred to our department. The patient had moderate myopia in both eyes (-5.00 D in the right eye and -4.50 D in the left eye). His medical history was otherwise unremarkable. The slit-lamp anterior segment examination and intraocular pressure measurement were normal; however, the ophthalmoscopic examination revealed optic disc edema and tilted disc in left eye (Figure 1). Peripapillary OCT demonstrated PHOMS in left eye that was located in the nasal sector (Figure 2, white arrows). Brain magnetic resonance imaging was unremarkable.

Case 2:

A 15-year-old boy presenting with suspected papilledema in left eye was referred to our department. The patient had moderate myopia in both eyes (-1.25 D in the right eye and -1.50 D in the left eye). His medical history was otherwise unremarkable. The slit-lamp anterior segment examination and intraocular pressure measurement were normal; however, the ophthalmoscopic examination revealed optic disc edema and tilted disc in left eye (Figure 3). Peripapillary OCT demonstrated PHOMS in left eye that was located in the nasal sector (Figure 4, white arrows). Brain magnetic resonance imaging was unremarkable.

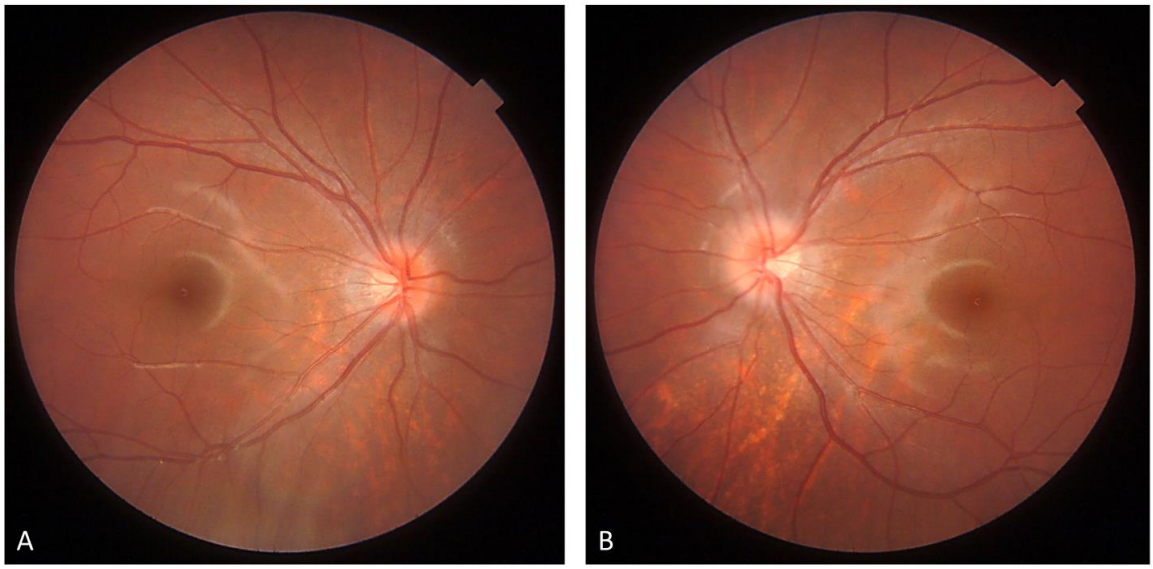

Fig 1: Fundus photographs of the right (A) and left (B) eyes

A pseudopapilledema can be observed in left eye because of the presence of tilted disc.

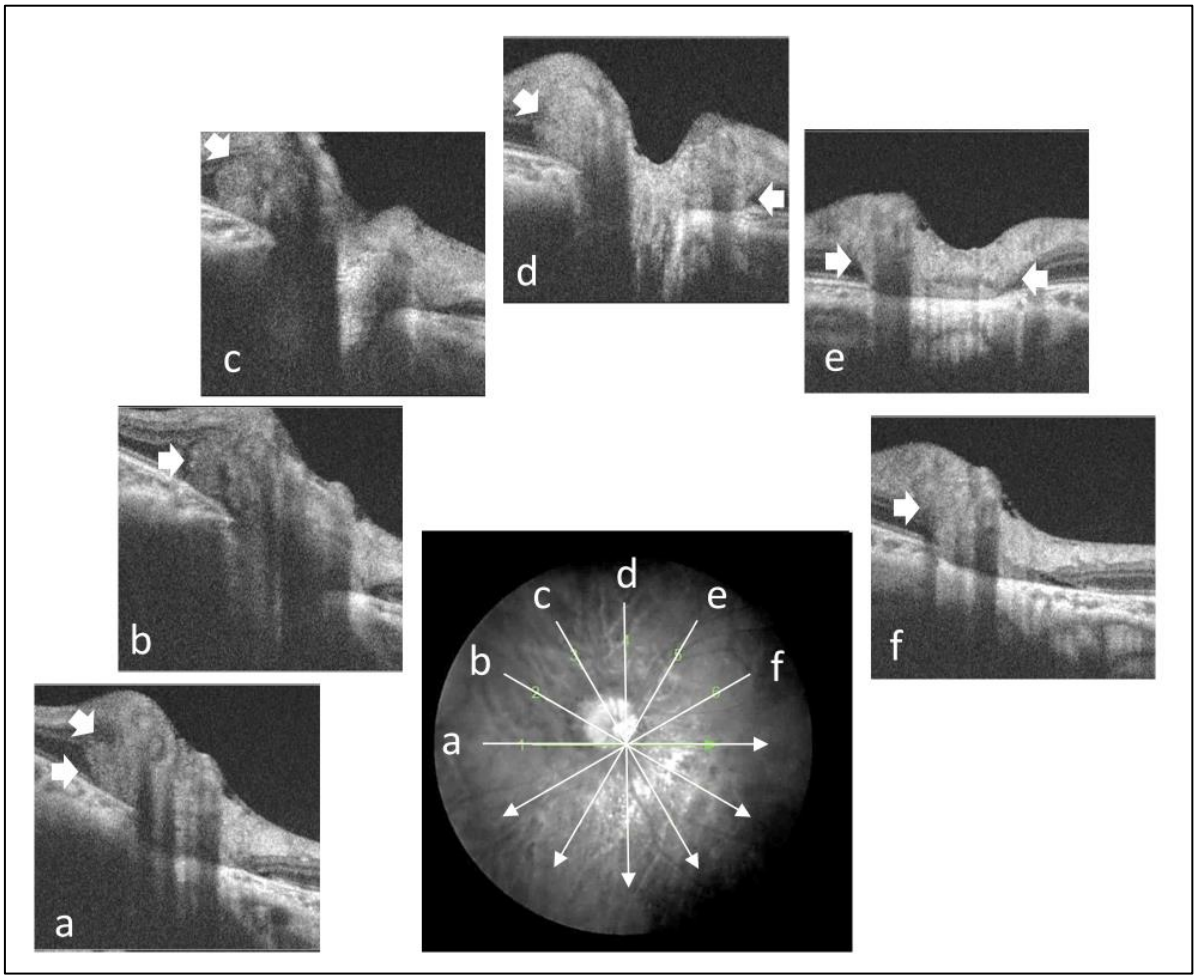

Fig 2: Optical coherence tomography of the left eye
Note hyperreflective ovoid mass-like structures that were located in the nasal sector (white arrows).

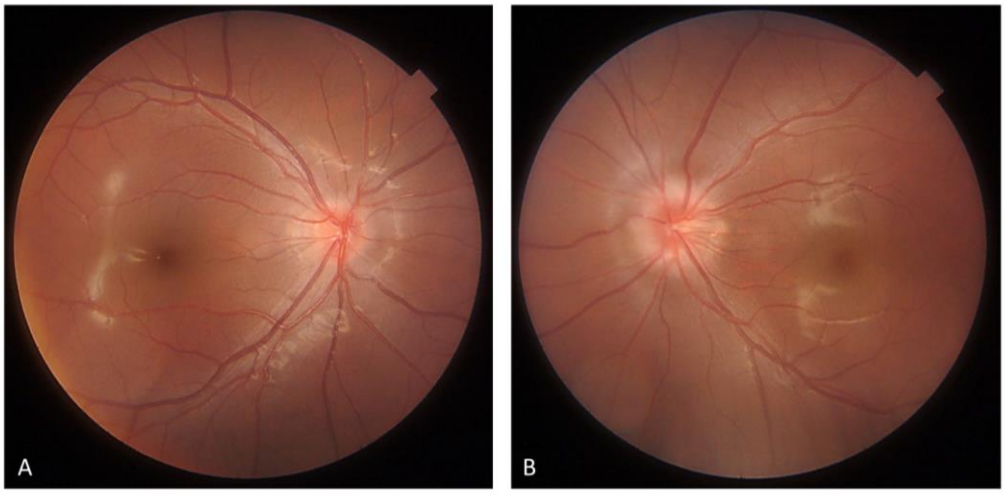

Fig 3: Fundus photographs of the right (A) and left (B) eyes

A pseudopapilledema can be observed in left eye because of the presence of tilted disc.

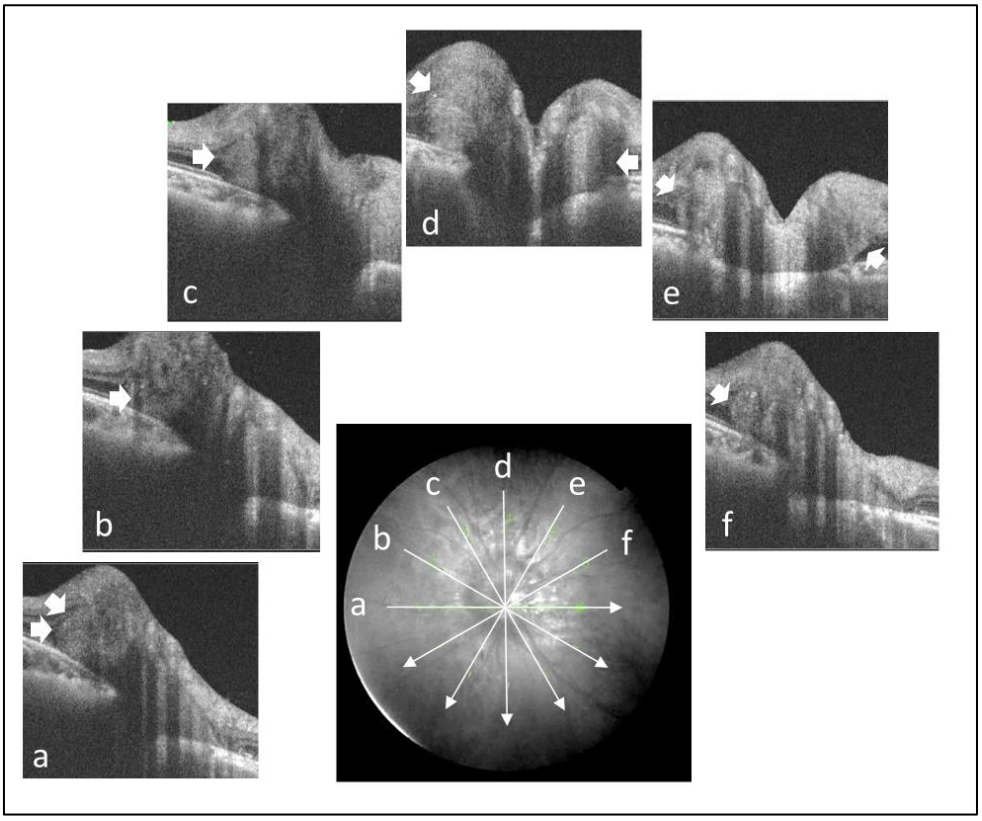

Fig 4: Optical coherence tomography of the left eye

Note hyperreflective ovoid mass-like structures that were located in the nasal sector (white arrows).

DISCUSSION

Based on these results, pseudopapilledema caused by PHOMS with tilted discs was diagnosed in present cases.

In an observational, population-based cohort study of 1,407 children aged 11–12 years, Behrens *et al*., [3] identified PHOMSs in 8.9% of patients. The PHOMS was predominantly found in the superonasal section of the optic disc. In addition, myopia and optic nerve head tilt were more common in children with PHOMS than in children without this condition. Pratt et al. [4] evaluated the OCT scans of the optic nerve in 110 children with suspected papilledema who were referred due to the presence of PHOMS, and reported the frequency of these structures. According to those authors, PHOMS were identified in at least one eye in 74 (67.3%) patients, with

42 (56.8%) of these showing bilateral and 32 (43.2%) displaying unilateral PHOMS.

Tilted disc syndrome (TDS) is an optic disc anomaly linked to myopia that manifests in a modest proportion (1–2%) of the general population [1, 2]. This high occurrence rate results in TDS causing a considerable level of pseudopapilledema in clinical settings, which is frequently associated with PHOMS [1, 2]. Lyu *et al*., [6] carried out a cross-sectional study that included 66 children with PHOMS and 46 control subjects, investigating the characteristics of children with PHOMS and evaluating associated risk factors. This comparative study identified the degree of myopia, and optic nerve head tilt angle as two notable risk factors for PHOMS.

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 [7]. 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 [8]. Thirdly, in pathological sections of optic neuritis models, Rao [9] found PHOMS, which was accompanied by demyelination of optic nerve fibers, extensive aggregation, and infiltration of inflammatory cells, leading to acute stasis of axoplasmic flow in nerve fibers of the optic disc. Fourthly, the nerve fiber damage in TDS cases is particularly evident at two sites: the entrance of the scleral canal and the level of the lamina cribrosa. Initially, Bruch's membrane projects toward the optic nerve at its superior extent, forcing the optic nerve fibers to bend sharply as they descend into the scleral canal. Subsequently, the lamina cribrosa is displaced relative to Bruch's membrane opening, and the gradual stretching of the sclera and lamina cribrosa may induce focal stress on optic nerve axons as they traverse the pores of the lamina cribrosa, particularly in the fibers that serve the nasal optic disc. This abnormal morphology can result in chronic axoplasmic stasis, which leads to nasal pseudopapilledema and optic disc elevation [5].

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. 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. Clinicians should be aware of PHOMS when a patient presents with myopic eyes and tilted disc syndrome, as in these cases.

REFERENCES

1. 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.
2. 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.
3. 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.
4. 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.
5. 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.
6. Lyu, I. J., Park, K. A., & Oh, S. Y. (2020). Association between myopia and peripapillary hyperreflective ovoid mass-like structures in children. *Scientific Reports*, *10*(1), 2238.
7. Skougaard, M., Heegaard, S., Malmqvist, L., & Hamann, S. (2020). Prevalence and histopathological signatures of optic disc drusen based on microscopy of 1713 enucleated eyes. *Acta Ophthalmologica*, *98*(2), 195-200.
8. Fraser, J. A., Sibony, P. A., Petzold, A., Thaung, C., Hamann, S., & Odds Consortium. (2021). Peripapillary hyper-reflective ovoid mass-like structure (PHOMS): an optical coherence tomography marker of axoplasmic stasis in the optic nerve head. *Journal of Neuro-Ophthalmology*, *41*(4), 431-441.
9. Rao, N. A. (1981). Chronic experimental allergic optic neuritis. *Investigative Ophthalmology & Visual Science*, *20*(2), 159-172.