—Case Reports—

Multimodal Imaging of Peripapillary Hyperreflective Ovoid Mass-Like Structures (PHOMS): Report of Two Cases

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Peripapillary hyperreflective ovoid mass-like structures (PHOMS) are a novel, incompletely understood finding in several optic nerve disorders. We describe PHOMS in two Japanese men. Both underwent general ophthalmologic examination as well as multimodal imaging, including visual field testing, fundus photography, spectral-domain optical coherence tomography (SD-OCT), OCT angiography (OCTA), B-scan ultrasonography, fundus autofluorescence (FAF) imaging, and fluorescence angiography. Case 1 was a 31-year-old man with normal pupillary reactions and normal visual acuity in both eyes. SD-OCT revealed PHOMS-oval hyperintense lesions—around the right optic disc. OCTA showed a vascular complex in the PHOMS, and FAF showed a blurry area of hypoautofluorescence at the optic disc. Case 2 was a 61-year-old man who, after undergoing several examinations, was diagnosed as having primary open angle glaucoma with a tilted disc. During a comprehensive examination, a color change was detected at the nasal side of the optic disc in his left eye; SD-OCT revealed PHOMS. One of these two Japanese patients with PHOMS had no other ophthalmic conditions, while the other had glaucoma with a tilted optic disc. Although SD-OCT and OCTA were essential in diagnosing PHOMS, multimodal imaging is required in order to rule out other disorders. (J Nippon Med Sch 2025; 92: 486–491)

Key words: peripapillary hyperreflective ovoid-mass like structures, spectral-domain optical coherence tomography, optical coherence tomography angiography, fundus autofluorescence

Introduction

Peripapillary hyperreflective ovoid mass-like structures (PHOMS) are an imaging finding that has been recently revealed after advances in enhanced depth imaging optical coherence tomography (EDI-OCT). PHOMS are not a precursor or subtype of optic disc drusen (ODD), but a separate OCT diagnosis[1]. One hypothesis is that PHOMS represent lateral herniation of distended retinal ganglion cell axons into the peripapillary region. The structures are wedged between the peripapillary nerve fiber layer and Bruch's membrane and form a torus or doughnut-like structure around the disc margin[2]. Previous studies reported an association between PHOMS and other ophthalmic conditions, including ODD[3–5], optic disc edema[6], idiopathic intracranial hypertension[7], non-arteritic anterior ischemic optic neuropathy[8], myopic/tilted optic disc[9], and multiple sclerosis-related optic neuritis[10,11]. However, Behrens et al.[12] reported that 8.9% of healthy children aged 11 to 12 years had PHOMS. Thus, it is likely that some adults also have PHOMS, without other optic diseases. Although PHOMS are present near the optic disc, few studies have investigated the relationship between PHOMS and glaucoma.

Other researchers have suggested that both EDI-OCT and optical coherence tomography angiography (OCTA) may have important roles in diagnosing PHOMS[13,14]. Although spectral-domain optical coherence tomography (SD-OCT) and OCTA are indeed crucial for diagnosing PHOMS, multimodal imaging examinations are necessary in order to rule out associated disorders.

In this study, we describe two cases of PHOMS in Japanese men who underwent multimodal imaging for other ocular disorders.

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Case Presentation

Both patients underwent general ophthalmologic examinations, including best-corrected visual acuity (BCVA), intraocular pressure, slit-lamp examinations, visual field and color fundus photography, as well as multimodal imaging, including SD-OCT, OCTA, B-scan ultrasonography, fundus autofluorescence (FAF) imaging, fluorescence angiography (FA), computed tomography (CT), and magnetic resonance imaging (MRI).

This case report was prepared in accordance with ethical guidelines for clinical research. Informed consent was verbally obtained from the patients for publication of this article and any images.

Case 1

A 31-year-old man with no relevant prior medical history was referred to our university hospital for further examination of an optic disc abnormality. He reported decreased vision in his right eye, for which he had sought treatment at a different clinic. His BCVA was 1.2 in both eyes, although his spherical equivalent was −0.75 in the right eye and 0 in the left eye. The difference in spherical equivalence between eyes was thought to be the cause of decreased vision in the right eye. Intraocular pressure was 13 mm Hg in the right eye and 14 mm Hg in the left eye, and the findings of a slit-lamp examination were normal. The central critical flicker fusion frequency was 37.3 Hz in the right eye and 40.7 Hz in the left eye. A fundus photograph revealed a slightly elevated mass lesion around the right optic disc, which changed color and resembled a papilledema. Tortuosity and dilation of the retinal artery was seen in both eyes (**Fig. 1A-1, 2**). SD-OCT showed round hyperreflective structures between Bruch's membrane and the retinal nerve fiber layer (RNFL) around the optic disc. The disc cup area was barely detectable (**Fig. 1B**). En face SD-OCTA of the same area revealed a vascular complex surrounding the optic disc (**Fig. 1C-1**). B-scan OCTA imaging showed a flow signal within the PHOMS (**Fig. 1D**). Both early-phase and late-phase FA showed staining without expansion at the PHOMS area (**Fig. 1E-1, 2**). FAF showed no hyperautofluorescence (**Fig. 1F**). The ganglion cell complex and RNFL thickness were normal (**Fig. 1G-1, 2**). Goldmann perimetry showed an enlarged blind spot in his right eye (**Fig. 1H-1, 2**), and B-scan ultrasonography confirmed the presence of a flat superficial hyperechoic structure without hyperechogenicity near the optic nerve head in his right eye (**Fig. 1I**). CT revealed a slightly hyperintense area, which was less bright than bone, on the optic disc (**Fig. 1J**). The patient had consulted a neurologist for a differential diagnosis, but no neurological abnormalities were found.

Case 2

During a routine annual physical examination, a 61-year-old man with no relevant prior medical history was suspected of having glaucoma and thus visited our university hospital. The patient had mild myopia in both eyes (−1.00 D in the right eye and −1.25 D in the left eye). BCVA was 1.2 in both eyes, and intraocular pressure was 13 mm Hg in the right eye and 14 mm Hg in the left eye. The findings of a slit-lamp examination were normal except for the presence of slight cataract in both eyes. The central critical flicker fusion frequency was normal and did not differ between the right and left eyes. A fundus photograph revealed an enlarged cup to disc ratio in both eyes, a nerve fiber layer defect on the superior and inferior nasal retina in his right eye (**Fig. 2 A-1, 2**), and topical retinal color change area at the nasal side of the optic disc in his left eye (**Fig. 2A-2**). SD-OCT showed hyperreflective structures between Bruch's membrane and the RNFL (**Fig. 2B**). En face SD-OCTA of the same area revealed a vascular network, but no vascular complex was visible (**Fig. 2C-2**). Additionally, SD-OCT showed thinning of the ganglion cell complex and RNFL in both eyes (**Fig. 2D-1, 2**). Visual field defects were detected in the right and left eye at the superior and inferior areas, respectively, as measured with the Swedish interactive thresholding algorithm (SITA) 30-2 standard automated perimetry (**Fig. 2E**). There was no indication of blind spot enlargement. He was diagnosed as having normal tension glaucoma (NTG) and prescribed eye drops.

Discussion

In this study, multimodal imaging was used to investigate PHOMS in one patient without any other disorders and in one patient with NTG and a tilted optic disc. Case 1 confirms that PHOMS may be present in patients without symptoms or other optic diseases. A previous population-based study found that 8.9% of healthy children aged 11 to 12 years had PHOMS and that myopia and optic nerve head tilt were more common in children with PHOMS than in those without PHOMS[12]. Lyu et al.[9] reported that myopia severity and optic nerve head tilt angle were two important risk factors for PHOMS. The present case 2 had a tilted disc in both eyes but only mild myopia. In contrast, case 1 exhibited neither myopia nor a tilted disc. Although there have been only two previously reported cases of PHOMS in Japanese[15,16], detec-

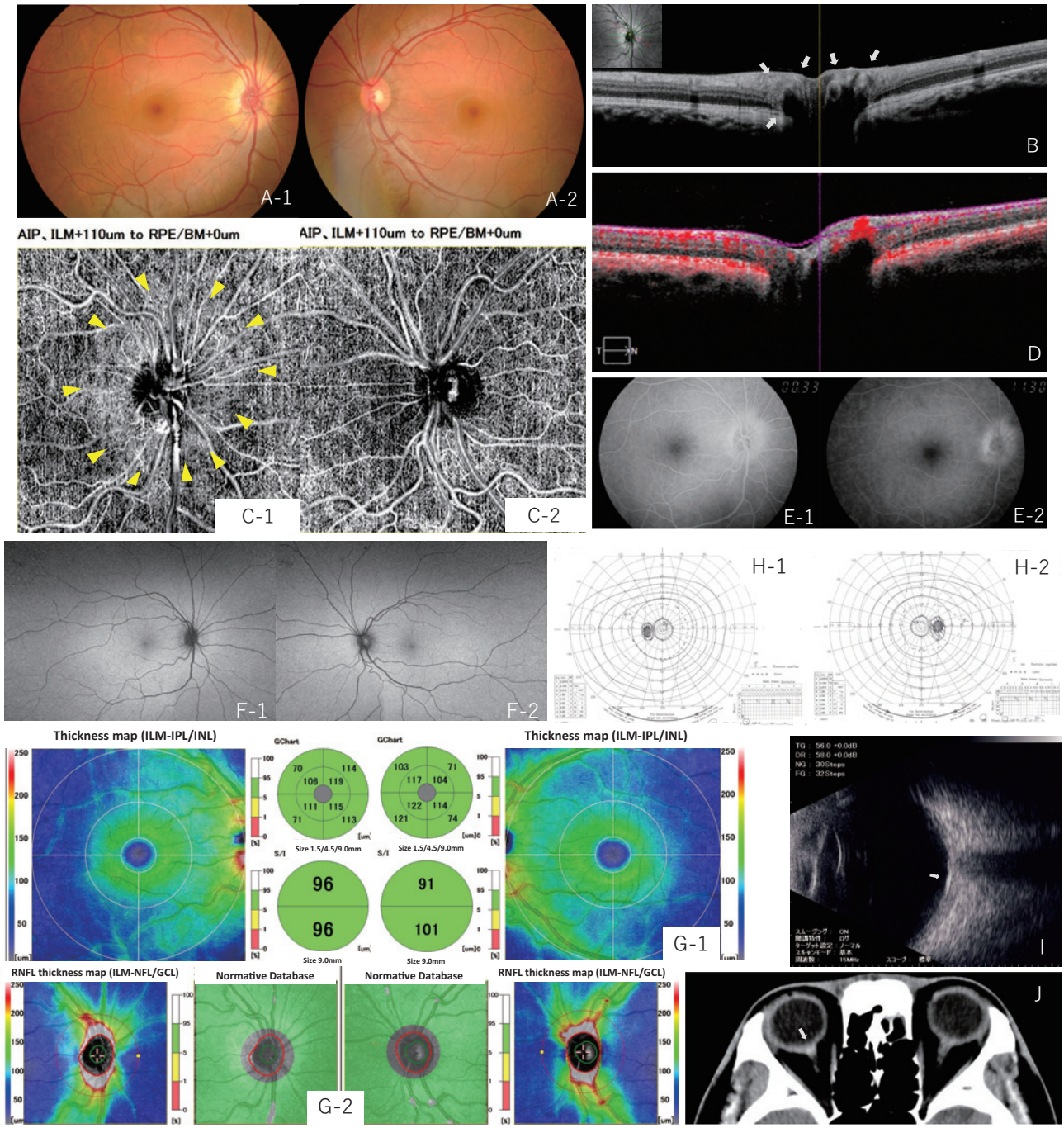

Fig. 1 Case 1: multimodal imaging

A fundus photograph shows a reddish optic disc surrounded by an annular mass in the right eye (A-1). The left eye appears normal (A-2). An SD-OCT image reveals PHOMS around the optic disc (white arrows) (B). This scan was obtained at the level of the red arrow on the near infrared image. En face SD-OCTA of the right eye shows a vascular complex around the optic disc (yellow arrowheads) (C-1). The vascular complex in the left eye is not as clear as that for the right eye (C-2). A flow signal is detected in the PHOMS (D). Fluorescence angiography shows fluorescence staining of PHOMS without expansion in both the early (E-1) and late (E-2) phases. Autofluorescence reveals an obscure boundary hypoautofluorescent area at the optic disc in the right eye (F). An SD-OCT image shows that the ganglion cell complex and retinal nerve fiber layer thickness were normal (G-1, 2). Goldmann perimetry shows an enlarged blind spot (H). B-scan ultrasonography of the right eye shows a flat superficial hyperechoic structure but no hyperechogenicity (I). CT shows an area of weak hyperintensity at the optic disc (J).

tion of PHOMS is likely to increase because of the high prevalence of myopia in Japan[17].

PHOMS was reported to be a cause of pseudopapilledema[1,6,18–20]. The incidence rate of PHOMS in patients with idiopathic intracranial hypertension was reported to be 62%[10,20]. In case 1, because a ring-shaped PHOMS around

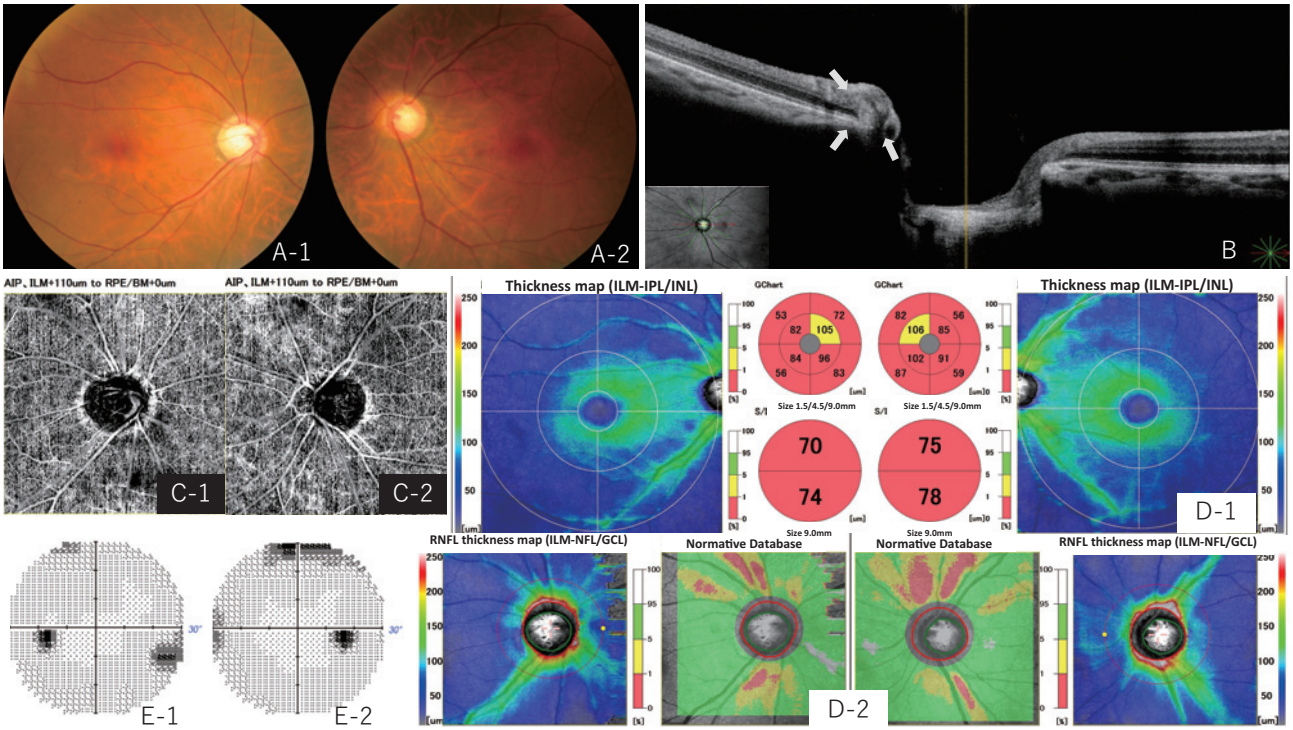

Fig. 2 Case 2: multimodal imaging

A fundus photograph shows a tilted disc and blurred boundaries at the nasal rim of the left optic disc (A-2). The right eye appears normal, except for an increased cup to disc ratio and NFLD (A-1). An SD-OCT image shows nasal PHOMS (white arrows) (B). This scan was obtained at the level of the red arrow on the near infrared image. The right eye showed no remarkable findings (C-1). En face SD-OCTA in the left eye shows the vessels of the PHOMS (C-2). SD-OCT showed thinning of the ganglion cell complex and retinal nerve fiber layer in both eyes (D). Humphrey Field Analyzer results for the left eye (E-1) and right eye (E-2).

the optic disc resembled pseudopapilledema, the patient underwent head CT and MRI. However, there were no findings associated with intracranial hypertension.

Borrelli et al.[13] reported that OCTA imaging revealed that PHOMS may contain a vascular complex. Additionally, Tsokolas et al.[21] reviewed OCTA findings in neurogenerative diseases, including multiple sclerosis, optic papilledema, and both arteritic and non-arteritic optic neuropathy. Previous studies reported that OCTA findings in multiple sclerosis, and in arteritic and non-arteritic optic neuropathy, showed a significant reduction in blood flow in the optic nerve head[22–24]. In contrast, a study of OCTA findings in papilledema found increased vessel tortuosity, density, and whole-image density[25]. In ODD, the vessel density of the optic disc was significantly diminished, particularly peripapillary capillary density[26,27]. OCTA may prove to be a valuable tool for differentiating optic disc diseases and monitoring disease progression. In our study, en face OCTA demonstrated the prevalence of vessels at the outer retinal level in both cases. The B-scan OCTA image in case 1 additionally showed a flow signal in the PHOMS. Long-term follow-up of this patient might reveal the origin of the vessels, whether the deeper vessels are displaced into the retina, and if there are new vessels that develop secondarily in the PHOMS.

The Optic Disc Drusen Study Consortium developed a consistent, standardized definition of ODD on OCT and introduced the distinct entity of PHOMS[1]. These lesions were described after reviewing 28 adults with proven ODD who were examined by EDI-OCT. Previous studies confirmed that EDI-OCT was more reliable than B-scan ultrasonography for ODD diagnosis. The authors of a previous study observed PHOMS in 28 out of 38 patients with ODD but were unable to identify PHOMS on ultrasound[1]. However, in our study, B-scan ultrasonography confirmed a flat superficial hyperechoic structure without hyperechogenicity near the optic nerve head in case 1. This suggests that ultrasound examination is important for distinguishing PHOMS from ODD, which is hyperechoic and represents hyperechogenicity. In our study, FAF analysis of PHOMS showed no hyperautofluorescence, whereas ODD showed hyperautofluorescence. These ultrasonography and FAF findings indicate that

these examinations are as important as OCT, especially when ruling out ODD.

Although some researchers maintain that PHOMS is not associated with RNFL loss[3], one of our patients had an enlarged blind spot on Goldman perimetry. During our evaluation of these patients, one image revealed that the RNFL was lifted by PHOMS. Thus, there could have been slight damage to the RNFL caused by this change. However, it would be difficult to detect if there were any morphological disc changes, as PHOMS are typically close to the disc. As for ODD, large, deep ODD might cause crowding and herniation of axons in the optic nerve head, thus leading to thickening of the superficial nerve fiber layer. Viana et al.[28] reported that patients with superficial ODD had a significantly lower visual field index, lower mean deviation, higher pattern standard deviation, thinner ganglion cell complex, and thinner RNFL than did patients with buried ODD. No reports have examined the relationship between the visual field and the presence of PHOMS. As seen in case 1, in which an enlarged blind spot was noted, PHOMS can cause visual field defects, depending on the size and the area involved.

The two present patients had no symptoms of disc-related disorders. Although cases of PHOMS in Japan are rare, we expect an increase in such cases because of the high prevalence of myopia in Japan[17]. The use of multimodal imaging, which includes EDI-OCT, FAF, and ultrasonography, is essential for characterizing PHOMS and determining the pathogenesis of nerve fiber herniation.

PHOMS alone is asymptomatic in the absence of complications. Thus, the present cases are rare, as PHOMS was detected fortuitously. Because PHOMS may be associated with several other disorders, patients with PHOMS should be followed for possible future complications.

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