We thank Lee et al. for their letter regarding our recently published recommendations regarding the diagnosis of optic disc drusen (ODD) (1). In the letter they raise several important questions.

The main purpose of our study was to establish well-defined recommendations for the diagnosis of ODD using OCT. During this process we came across the peripapillary hyperreflective mass-like structures (PHOMS), which are discussed in the letter. Several studies have previously diagnosed PHOMS as buried ODD (2-5). We argue that PHOMS are not ODD but are in fact herniating nerve fibers based on the observations that:

1) The morphology of PHOMS on OCT differs from ODD that have been verified by fundus photography or autofluorescence (6).

2) Histopathology from eyes with papilledema stain positive for nerve fibers in areas corresponding to PHOMS (7).

3) Histopathology of PHOMS from eyes with ODD is indistinguishable from PHOMS seen in eyes with papilledema (8).

4) PHOMS are always located in the peripapillary circumference, corresponding to the area of optic disc margin blurring in patients with papilledema or pseudopapilledema (1).

5) PHOMS are found in a variety of conditions with optic disc swelling such as idiopathic intracranial hypertension (IIH) (9).

6) We have evidence that PHOMS regress in IIH patients after intracranial pressure (ICP) lowering therapy (unpublished data).

That PHOMS should be diagnosed as herniating nerve fibers above the Bruch's membrane layer is also supported by Wang et al. in a recently published study (6).

We agree with Lee et al. that PHOMS have only been possible to visualize due to emerging OCT techniques, but as PHOMS are located close to the inner limiting membrane, the use of enhanced depth imaging is not a requirement. Further, we agree with Lee et al. that PHOMS should be regarded as a different entity. We believe it is a non-specific marker of distended or displaced axons, axoplasmic stasis or congestion in the prelaminar optic nerve head. In patients with papilledema the axoplasmic stasis is most likely caused by increased retrolaminar tissue pressure induced by high ICP. In ODD patients with pseudopapilledema, PHOMS is most likely caused by local congestion, possibly due to a primary axonopathy or mass effect compressing neighboring axons, although the specific pathophysiology is still not understood.

When it comes to the diagnosis of ODD using OCT, we don't agree with Lee et al. that we need to restrict our definition of ODD to include only calcified masses within the optic nerve head. In our study we also included buried ODD. These ODD were situated deep in the optic nerve head and were hyporeflective, with minimal or no hyperreflective margin. In 5-year follow-up EDI-OCT scans from a previously published study of children with ODD (10), we saw that hyporeflective areas in the deep layer of the optic nerve head (what we define as buried ODD) evolved into larger and slightly more calcified ODD (Fig.1). However, when looking through the follow-up data we never saw PHOMS evolve into ODD (Fig. 2).

Could PHOMS be an early form of ODD or turn into ODD as speculated by Lee et al. and previously by Traber et al. (4)? We too, have seen ODD located inside PHOMS, but why wouldn't they? ODD can be found everywhere among the axonal tissue of the prelaminar optic nerve head, and there is no reason not to expect them within the nerve fibers of PHOMS as well. We believe that there is currently no evidence to support the theory that PHOMS are early ODD or might evolve into ODD.

As we argue above, PHOMS should not be seen as a differential diagnosis of optic disc edema but rather as a manifestation of the edema. PHOMS do not always correlate to the degree of papilledema or pseudopapilledema and sometimes we see PHOMS in individuals with healthy eyes (Fig. 2). Most likely, hereditary optic disc anatomy and other factors play a role in the development and degree of PHOMS, rather than being an independent disease process.

By recognizing PHOMS as a marker of axoplasmic stasis, we might be able to use PHOMS in the diagnostics and disease tracking of different optic neuropathies. Only future studies will tell us the possible impact of this finding, but in the meanwhile we would caution against confusing PHOMS with early or buried ODD, even though they may sometimes coexist.

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