Coexistence of Paracentral Acute Middle Maculopathy and Prominent Middle Limiting Membrane in Purtscher-like Retinopathy

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ABSTRACT

Purtscher-like retinopathy is a rare clinical entity that develops due to non-traumatic causes, in which ischemia secondary to micro-vascular alteration is implied in the pathogenesis. The paracentral acute middle maculopathy is one of the optical coherence tomography findings of acute retinal ischemia and indicates deep capillary plexus ischemia, which is characterized by a hyperreflective band in the middle retinal layers in the acute phase. Prominent middle limiting membrane sign, another optical coherence tomography finding of retinal ischemia, is observed as a hyperreflective line in the inner aspect of outer plexiform layer. Herein, we report a patient with the signs of Purtscher-like retinopathy secondary to systemic hypertension and discuss accompanying paracentral acute middle maculopathy and prominent middle limiting membrane findings. With this report, it was aimed to emphasize the novel findings of acute retinal ischemia that can be detected with clinical examination and imaging methods.

Key words: Optical coherence tomography, optical coherence tomography angiography, paracentral acute middle maculopathy, prominent middle limiting membrane, Purtscher-like retinopathy.

INTRODUCTION

The Purtscher retinopathy is a rare entity that was first defined in 1910 and develops following traumas which does not directly involve eyes. The Purtscher-like retinopathy is defined as clinical entity that occurs secondary to non-traumatic causes. The primary causes leading Purtscher-like retinopathy include acute pancreatitis, connective tissue disorders, chronic renal failure, malignancies, hemolytic uremic syndrome, orbital injections, cryoglobulinemia, Valsalva maneuver and delivery. The most common clinical findings are cotton wool appearance, areas of inner retinal whitening (Purtscher flecken) and hemorrhages. Although pathogenesis is controversial, it is thought that these findings result from retinal ischemia secondary to embolic occlusion of pre-capillary arterioles[1, 2].

The paracentral acute middle maculopathy (PAMM) is a novel finding of acute retinal ischemia which is thought to be due to deep capillary ischemia and identified by hyper-reflective band appearance in middle retinal layers (particularly in inner nuclear layer) on optical coherence tomography (OCT)[3]. It often accompanies to retinal vascular occlusion[4-6]. Recently, a prominent middle limiting membrane (MLM) finding was defined, which is considered as a marker of acute retinal ischemia in retinal vein occlusions and observed as a hyper-reflective line at synaptic part of outer plexiform layer (OPL)[7, 8].

Here, we presented a case of Purtscher-like retinopathy with PAMM and prominent MLM.

CASE REPORT

A 48-years old woman presented with decreased vision in left eye over 3 days. There was no history of trauma or known disease other than uncontrolled hypertension. In the history, it was found out that she was given anti-hypertensive treatment with systemic blood pressure of 200/120 mmHg one week before presentation. In the ophthalmological examination, the best-corrected visual

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Received: 14.05.2020
Accepted: 05.06.2020

Ret-Vit 2021; 30:303-307

DOİ: 10.37845/ret.vit.2021.30.53

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acuity was 20/20 in right eye and 20/100 in the left eye. Intraocular pressure was normal in both eyes and anterior segment examination was found to be normal. In the fundus examination, thinning in retinal arteries was observed in both eyes; in addition, it was seen that there was cotton wool appearance at peripapillary region and lobular intraretinal whitening involving fovea at macula in the left eye (Figure 1). On fluorescein angiography, no vascular occlusion or abnormal staining was observed (Figure 1). In left eye, there was a hyper-reflective band appearance passing across areas of intraretinal whitening with skipping pattern at inner nuclear layer on OCT, which is characteristic for PAMM[3]. In addition, a prominent hyper-reflective line (MLM) was detected at inner aspect of outer plexiform layer (Figure 2). On en-face OCT angiography (OCTA), a perivenular fern-like appearance at deep capillary plexus and areas of hypo-perfusion in deep capillary plexus were striking, which are well-known findings in PAMM (Figure 2)[9, 10]. The imaging findings were considered as normal in the right eye. The patient was referred to internal medicine and cardiology departments for further evaluation due to Purtscher-like retinopathy and concurrent findings of acute vascular ischemia (PAMM and prominent MLM). No systemic disease other than hypertension was detected in the patient. No change in BCVA was detected in the control visit on week 6. In fundus examination, it was observed that peripapillary cotton wool appearance was resolved in left eye. On OCT, it was observed that the hyper-reflective band appearance at INL and prominent MLM was resolved; however, there was marked thinning in INL (Figure 3). On en-face OCT angiography, perivenular fern-like appearance observed in acute period was resolved with shrinking in areas of hypo-perfusion at deep capillary plexus (Figure 3).

DISCUSSION

Although the pathogenesis in Purtscher-like retinopathy hasn't been fully elucidated, it is thought that the entity develops as a result of leukocyte aggregation due to complement activation caused by precursor events. The leukocyte aggregates formed lead occlusion in retinal arterioles and secondary infarction[1, 2]. In systemic hypertension, it was shown that free oxygen radicals released led oxidative stress and impaired end-organ micro-circulation together with vascular endothelial injury, platelet activation and aggregation[11]. In our patient, the only etiological factor identified was hypertension which might have caused Purtscher-like retinopathy by leading pre-capillary arteriolar occlusion. The cotton wool appearance is observed when pre-capillary arteriolar occlusion involved superficial capillary plexus while PAMM develops when it involved deep capillary plexus.

PAMM was first defined as a variant of acute macular neuroretinopathy by Sarraf et al. In 2013[3]. However, it was shown that it is a distinct clinical finding caused by ischemia in deep capillary plexus of retina, rather than being a specific retinal disease, in subsequent publications.

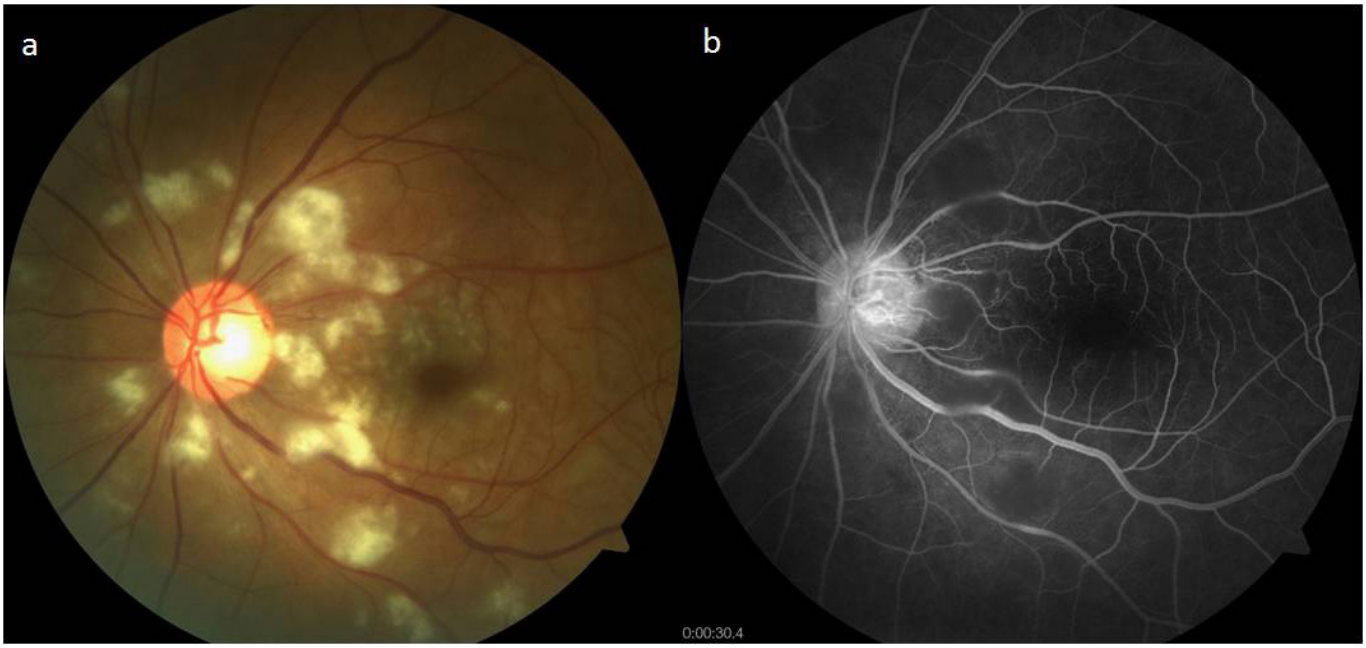

Figure 1: a) *cotton wool appearance at peripapillary region and areas of lobular intraretinal whitening involving fovea at macula are observed on color fundus image of left eye*. **b)** *blocked areas are seen due to cotton wool appearance but no vascular occlusion or abnormal staining on fluorescein angiography.*

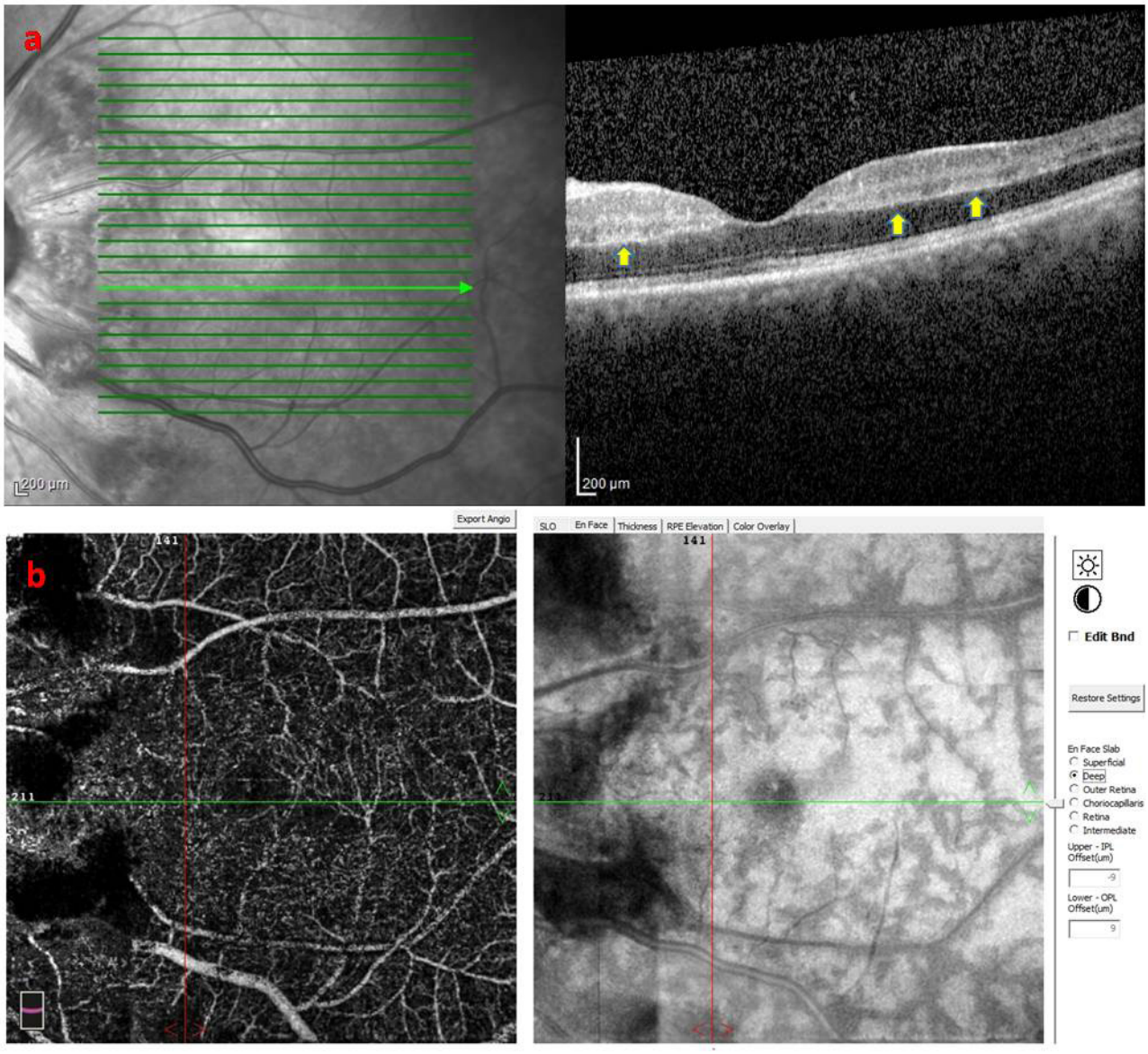

Figure 2: a) *Hyper-reflective band appearance (PAMM) with skipping pattern at inner nuclear layer and marked hyper-reflective line (prominent MLM) at inner aspect of outer plexiform layer are observed on OCT imaging of left eye (yellow arrows);* **b)** *in same eye, perivenular fern-like appearance is seen in the Figure on right and areas of hypo-perfusion on deep capillary plexus in the Figure at left in the en-face OCT angiography.*

In recent publications, it was reported that PAMM is associated with conditions affecting retinal micro-circulation such as retinal artery and vein occlusions, diabetic retinopathy, hypertensive retinopathy, Purtscher retinopathy and sickle cell retinopathy[4-6]. On OCT, acute lesions of PAMM are seen as hyper-reflective bands at level of INL; followed by INL thinning and atrophy which are observed as disorganization. On OCTA, PAMM is characterized by areas of hypo-perfusion at deep capillary plexus[10]. Three patterns have been defined for PAMM on en-face OCT images: arteriolar, globular and fern-like (perivenular) appearance[9]. The fern-like appearance has been solely reported in cases secondary to central retinal vein occlusion (CRVO) so far. Moreover, fern-like appearance on en-face OCT is considered in favor of CRVO in cases where clinical presentation is vague. It is though that perivenular PAMM results from arterial failure due to CRVO[12]. In our case, it is seen that primary arterial hypo-perfusion, albeit extremely rare, may lead perivenular PAMM.

The prominent MLM sign, which was first described by Chu et al. in 2013, is considered as OCT findings of acute retinal ischemia and indicates poor visual prognosis[7, 8]. Although it can be seen at acute phases in retinal artery occlusions, it accompany to CRVO. The prominent MLM manifesting as a hyper-reflective line at inner aspect of outer plexiform layer results from cytoplasmic swelling due

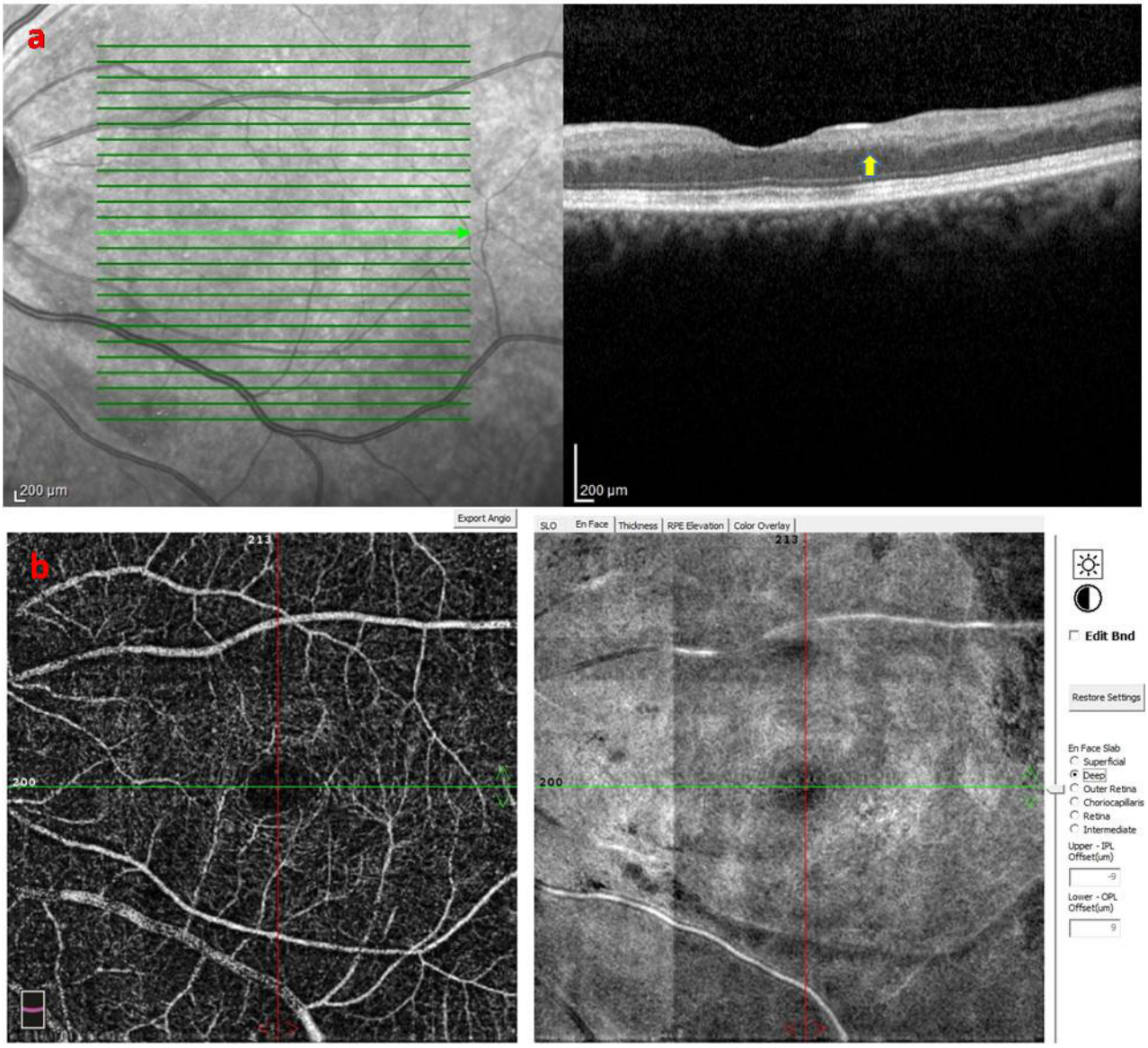

Figure 3: a) *It is seen that hyper-reflective band image at inner nuclear layer and prominent MLM are resolved but inner nuclear layer is thinner and disorganized on OCT imaging on week 6 (yellow arrow).* **b)** *in same eye no perivenular fern-like appearance is observed in the Figure at right while areas of hypo-perfusion at deep capillary plexus are reduced in the Figure at left in en-face OCT angiography.*

to ischemia in bipolar cells but not a true membrane. The outer plexiform layer is outermost later of retinal vascular supply with high oxygen demand; thus, it is susceptible to alterations in retinal perfusion.

In the literature, a case series was reported, including 3 patients with central retinal artery occlusion presenting with Purtscher-like retinopathy[13]. In all three cases, cotton wool appearance and cherry red spot were observed with prolonged branch-retinal circulation time on fluorescein angiography. In an experimental study, it was shown that an embolism of 150 µm can lead retinal finding similar to Purtscher retinopathy. However, clinical and imaging findings excluded central retinal artery occlusion in our case.

In conclusion, Purtscher-like retinopathy is a rare clinical entity with unknown etiology. PAMM and prominent MLM sign at acute phase may be considered as biomarkers of deep capillary ischemia and are associated with poor prognosis.

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