Isolated Sub-internal Limiting Membrane Fluid Accumulation as a Rare Manifestation of Optic Disc Pit Maculopathy

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ABSTRACT

In this article, it was aimed to demonstrate sub-ILM fluid accumulation, one of the findings of optic disc pit maculopathy, in a 16-year-old male patient with refractive error using optical coherence tomography. The presence of sub-ILM fluid reported in optic disc pit maculopathy is usually associated with fluid accumulation in the intraretinal or subretinal area, and isolated sub-ILM fluid accumulation is a rare finding that should be emphasized.

Keywords: Internal limiting membrane, Maculopathy, Optic disc pit.

INTRODUCTION

Optic disc pit was first described by Wiethe in 1882, which is a congenital anomaly manifested as grayish, limited depression at lamina cribrosa of optic disc.[1,2] Approximately 70% optic disc pits are localized at temporal to optic disc while 20% at central and remaining at nasal to optic disc.[3] Macular problems occur eventually and visual prognosis is poor.[4] Maculopathy accompanied by serous detachment and retinoschisis is more frequent when pit is localized at temporal and larger.[2] Although origin and pathophysiology of fluid remain unclear, it is thought that it originates from vitreous or cerebrospinal fluid. Here, it was aimed to present a case with optic pit accompanied by fluid accumulation under internal limiting membrane (ILM) which was followed over 2 years.

CASE REPORT

A 16-years boy presented with difficulty in distant acuity. In the ophthalmological examination of the patient, visual acuity was 20/25 bilaterally without correction and 20/20 with -0.75 axis 180 refraction correction. Intraocular pressure was 14 mmHg in both eyes. Biomicroscopic anterior segment examination was normal. In fundus examination, there was optic disc pit at inferotemporal to optic disc and irregularity in retinal nerve fiber layer (RNFL) at same quadrant in the right eye while macula was normal (Figure 1a). No abnormal finding was observed in the left eye. Optical coherence tomography (OCT) imaging showed fluid accumulation under the ILM (Figure. 2) and hypo-autofluorescence due to fluid under the ILM was observed in the related area (Figure. 3).The patient was diagnosed with optic disc pit and secondary sub-ILM fluid and followed over 2 years. During follow-up, it was seen that visual acuity was preserved and sub-ILM fluid persisted with minimal fluid under ganglion cell layer on OCT (Figure 4). In SITA Faster visual field testing on year 2, enlargement of blind spot compatible with RNFL changes on OCT was observed (Figure 5)

DISCUSSION

Optic disc pit is a rare congenital anomaly which is thought to occur due to failure of fetal fissure closure during embryogenesis. In histopathology, it was shown that dysplastic retina is herniated into subarachnoid space

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Received: 30.11.2022
Accepted: 26.12.2022

J Ret-Vit 2023; 32: 276-279

DOİ:10.37845/ret.vit.2023.32.47

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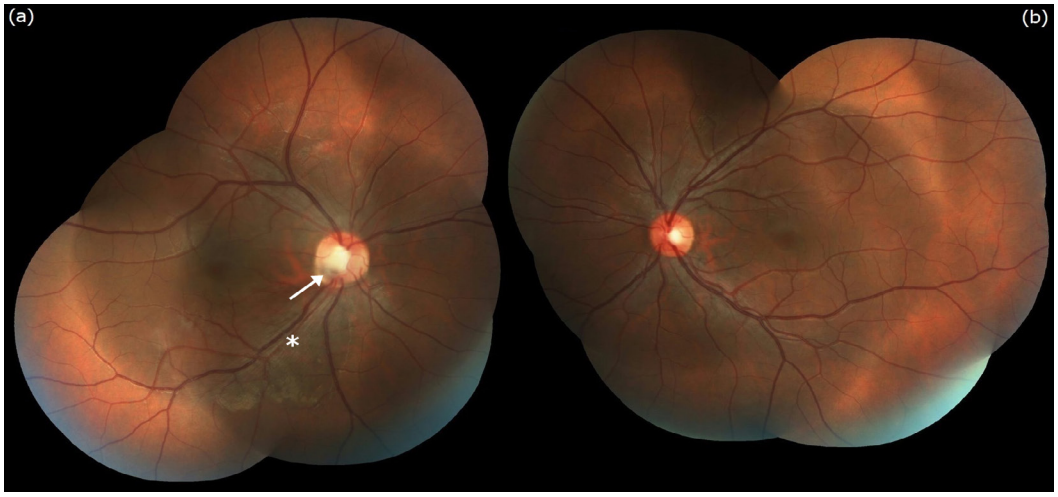

Figure 1: *a) Color fundus image of right eye. Pit at inferotemporal to optic disc (white arrow) and irregularity in inferotemporal retinal nerve fiber layer (*) are seen. b) Color fundus image of left eye. Normal optic disc and macula are seen.*

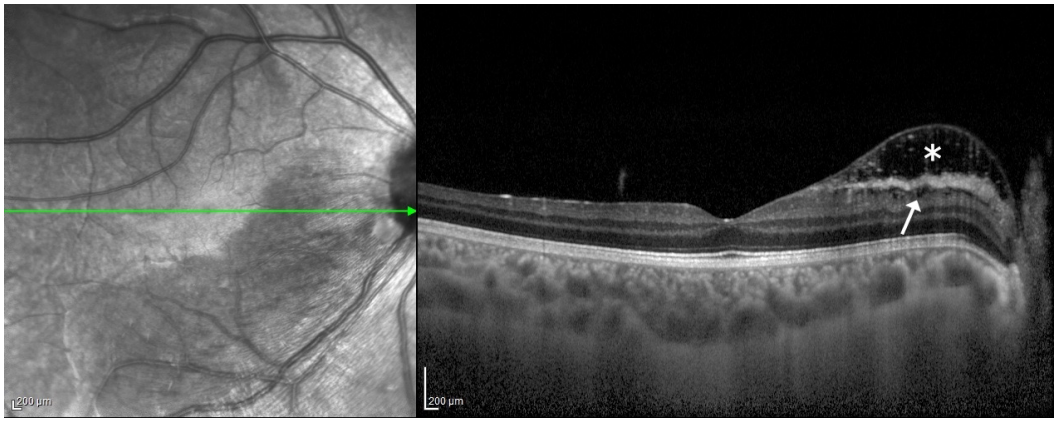

Figure 2: *Optical coherence tomography image of right eye. Fluid forming schisis-like cavitation/elevation under internal limiting membrane (*). Somewhat amount of fluid extends under ganglion cell layer (white arrow).*

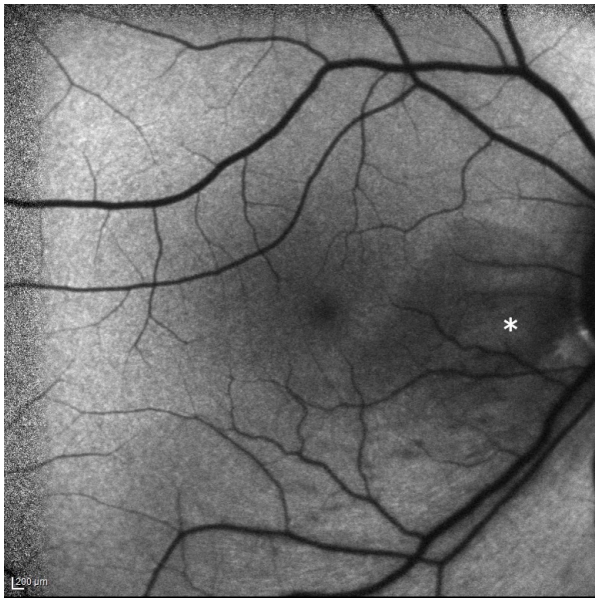

Figure 3: *Autofluorescence of right eye. Hypo-autofluorescent appearance due to fluid under internal limiting membrane (*).*

through lamina cribrosa. The estimated prevalence is 2:10,000 without gender preponderance. Although it is typically unilateral, bilateral presentation was reported in 15% of cases.[5]

The patients with optic disc pit are asymptomatic unless complication develops; however, 25-75% of patients will experience optic disc pit maculopathy characterized central serous macular detachment at some point in their life.[3,6] There is no consensus on origin of subfoveal fluid; in addition, the mechanism underlying fluid accumulation hasn't been clearly elucidated. It is thought that fluid originates from vitreous or cerebrospinal fluid in optic disc pit maculopathy without subretinal fluid leakage on fluorescein angiography. Lincoff et al. proposed that fluid from optic disc pit initially leads schisis-like formation at inner retina; which, then, moves into subretinal area through hole at outer layers.[7] However, by introduction of OCT, it was reported that outer nuclear layer is the most

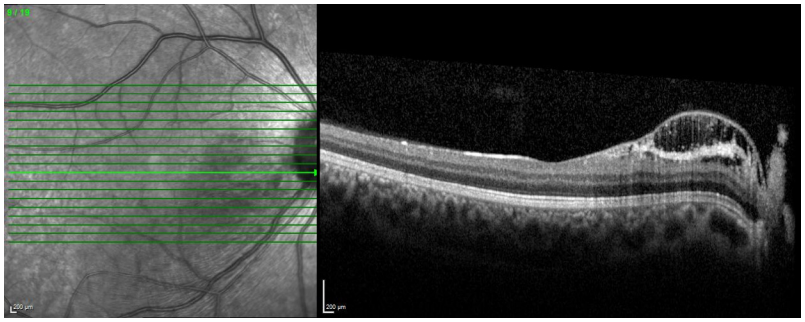

Figure 4: *Optical coherence tomography image of right eye. In control visit on year 2, minimal fluid increase under internal limiting membrane and ganglion cell layer.*

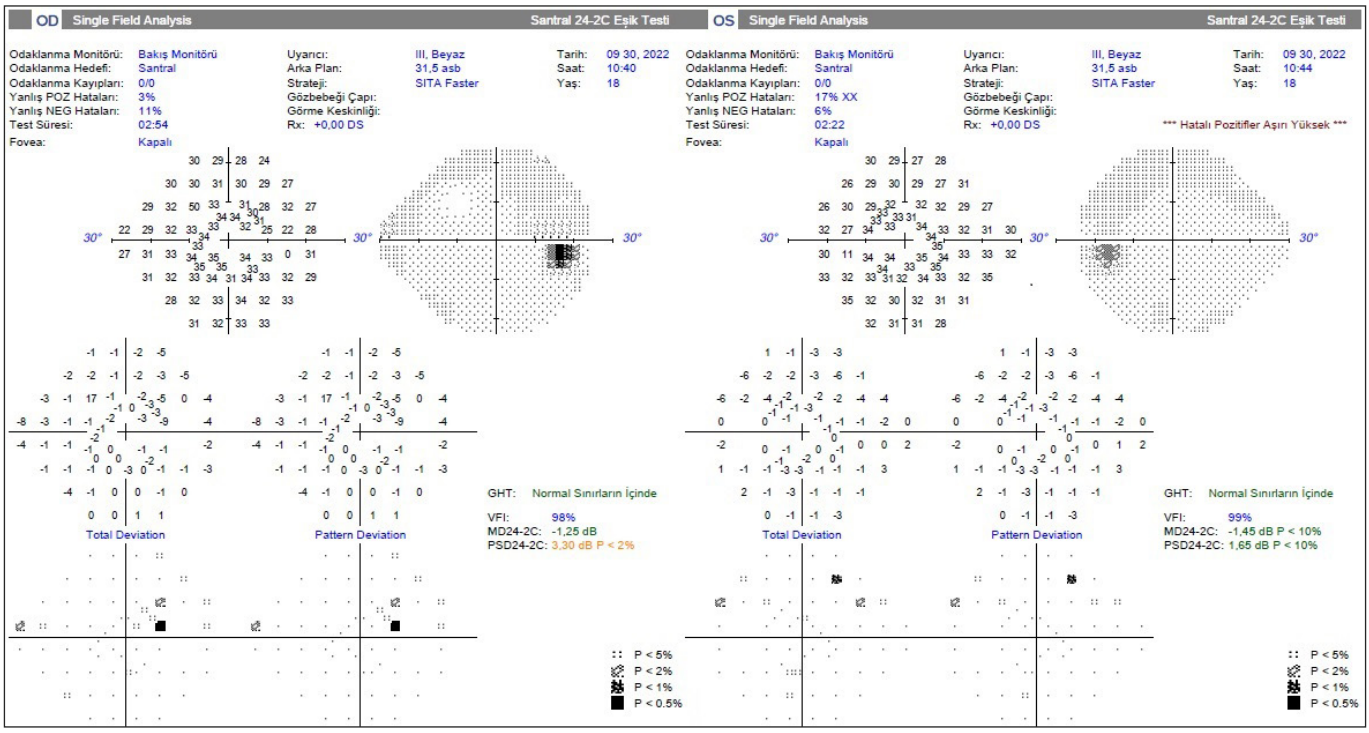

Figure 5: *SITA Faster visual field testing in right and left eye. Enlargement of blind spot compatible to RNFL changes in right eye.*

severely affected region and that the fluid could directly move under ILM and to ganglion cell layer, inner nuclear layer or subretinal area.[8]

The sub-ILM fluid in optic disc pit maculopathy was generally shown together with fluid accumulation in other retinal layers or subretinal area. Optic disc pit progressing with isolated fluid accumulation under ILM is extremely rare. As it was the case in our patient, it should be kept in mind that schisis-like cavitation/elevation appearance in association with sub-ILM fluid may be present without subretinal fluid or retinoschisis extending up to fovea. It is important to be aware of such association in diagnosis and follow-up of optic disc pit.

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