The Clinical Ophthalmologist Journal

ISSN: 2692-7985

Case Report

Pseudo-Tripling of Optic Disc: A Case Report

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Abstract

Background: Although the coloboma of the optic nerve head and/or chorioretinal tissue is not uncommon, duplication or pseudo-duplication of the optic disc is rare. Here we report a rare case of pseudo-tripling of optic disc, in a 50 years-old asymptomatic man. Orbital MRI confirmed presence of only one optic nerve.

Keywords: Optic disc doubling; Optic disc coloboma; Chorioretinal tissue

Introduction

Doubling of the optic disc is a rare clinical entity. Pseudo-doubling of the optic disc is also relatively uncommon, and there are only a few reports of this finding. Pseudo-duplication of the optic disc is considered to be generally associated with chorioretinal colobomas. Differentiating between these two entities might face several challenges. Our case represents a unilateral pseudo-tripling of the optic disc. Up to our knowledge this is the first case of such a finding in the literature.

Case Presentation

A 50 years-old man is referred to the ophthalmology clinic by his primary care physician for evaluation of possible diabetic retinopathy. He denies any visual problem rather than near vision difficulty, for which he was wearing near glasses since several years ago. His past medical history was positive for newly diagnosed type 2 diabetes. His family history was negative for any ophthalmic disease.

The patient underwent full ophthalmic examination. The best corrected visual acuity was 10/10 in both eyes. There was no Relative Afferent Pupillary Defect (RAPD). The slit lamp examination revealed nothing but an early cataract in both eyes. The intraocular pressure was 15 mmHG in both eyes. The fundoscopy of right eye revealed a medium-size disc with a cup to disc ratio of 0.3, and a few pigmentary changes in nasal macular region. No diabetic retinopathy was detected (Figure 1). In the fundoscopy of the left eye, the optic disc was relatively smaller than the right eye, with cup to disc ratio of 0.2. Just in the inferior part of the optic disc, with a distance of 0.5 disc diameter, there is an oval-to-round shape lesion, with approximately half of the size of the disc, resembling to a "second" optic disc. There was an area of chorioretinal atrophy, just superior to this lesion. The major vessel of the inferior arcade crossed this "second" optic disc and going on its way to reach a third lesion, just inferior to the second optic disc. The latter is a round, white lesion with size of approximately three optic discs. This "third" pseudo-optic disc lesion seemed to be a chorioretinal coloboma. No diabetic retinopathy was also detected in this eye (Figure 2). A Magnetic Resonance Imaging (MRI) scan of the orbits revealed just one optic nerve on each side.

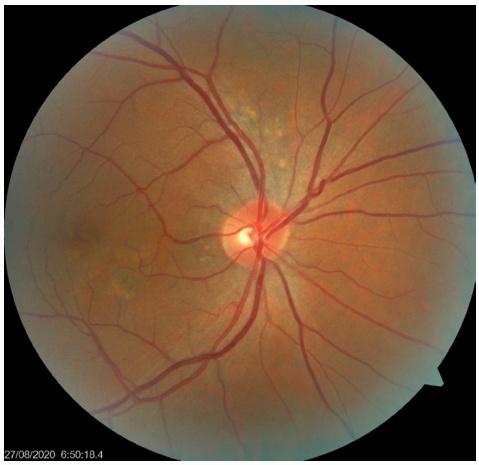

Figure 1: Fundus photography.

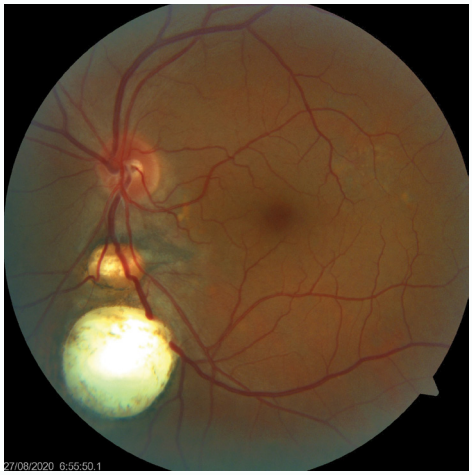

Figure 2: Fundus photography.

Citation: Farzinvash Z. Pseudo-Tripling of Optic Disc: A Case Report. Clin Ophthalmol J. 2021;2(1):1017.

Publisher Name: Medtext Publications LLC

Manuscript compiled: Feb 10th, 2021

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We assure the patient that this finding was not related to diabetes and will not go to harm visual function. After discussion with the patient, written informed consent was taken to use the fundus photography for publication purpose. The patient was scheduled to be examined for diabetic retinopathy after one year.

Discussion

Both duplication and pseudo-duplication of the optic disc are rare clinical entities. The typical presumed "second" optic disc is usually a circumscribed, optic disc-like lesion, located 0.5-3 disc diameters inferior to the actual optic disc, show apparent cupping and usually associated with surrounding chorioretinal atrophy. The central retinal vessels are bridging the lesion and there are vessels which are radiating from the area [1,2]. Pseudo-duplication is generally thought to be associated with coloboma [3-6]; but there are case reports in which this condition has been reported with some other conditions, for example high myopia [7], CHARGE syndrome [8], or proliferative diabetic retinopathy [9]. True duplication of the optic nerve is extremely rare. According to a case report by Brink et al. [3], Elsching in 1914 described a case of doubling of the optic nerve, identified in autopsy. Lamba described bilateral coloboma of the choroid and doubling of the left optic disc together with presence of two optic foramina, observed by X-ray examination [10]. Pesme (1948) and Kubik (1925), cited by Brink et al. [3], reported cases of double optic discs with independent vascular supplies.

The differentiation of true double optic disc from pseudo-doubling can be challenging; the true second disc must fulfill one of the findings of extra nerve fiber layer, separate vascular supply, or presence of two separate optic nerves on MRI or CT scan of the orbit [3,10]. Although in a case report by Padhi et al. [11], the second optic disc had separate vasculature but there was only one optic nerve shadow in B-scan of the globe.

It is worth mentioning that in any case of optic disc doubling, the first diagnosis to be ruled out is optic head nerve and chorioretinal colobomas. A typical coloboma occurs due to closure defects in the proximal embryonic fissure at six weeks of gestation, and optic nerve head and chorioretinal colobomas can occur together [4,6,9,12]. A precise fundoscopic examination can differentiate true doubling from more common diagnosis of optic disc pseudo-doubling. If in doubt, paraclinic evaluation can help; Fluorescein angiography can show the separate vasculature of two optic discs in the case of true doubling [2,5,11-13]. B-scan ultrasonography can reveal the shadow of extra optic nerve, and CT-scan or MRI of the orbit can confirm the diagnosis. Perimetry can show double blind-spot, but this finding may also be seen in the case of pseudo-doubling [2,5]. Most colobomas occur in the inferior part of the optic disc, so superior visual field defects may be seen in pseudo-doubling [5,11]. Even colour Doppler imaging study can help to differentiate these entities [12]. Sometimes pseudo-doubling of the optic disc encounter with other ocular conditions, such as macular congenital hypertrophy of the RPE [5], macular schisis [13], or bilateral optic disc pits [14].

Here we present a case of pseudo-tripling of the optic disc. In this case, there is a lesion resembling second optic nerve. This lesion had its vascular supply from the inferior major arcade of the original optic nerve, with surrounding chorioretinal atrophy; although it resembles a "second" true optic disc regarding its shape, color, and cupping, it seems to be an optic disc coloboma in origin. The third lesion which is below the "second" optic disc is apparently a chorioretinal coloboma. Orbital MRI confirmed the presence of one optic nerve in the left orbit. As to our knowledge, this is the first report of such a finding in the literature.

In conclusion, hereby we present a case of unilateral pseudo-tripling optic disc in an asymptomatic man. Our case doesn't have any other ocular abnormality. True doubling of optic nerve head is a rare clinical condition. It can be differentiated from pseudo-doubling by the means of delicate fundoscopy and/or paraclinic evaluation. Both conditions usually cause no harm to visual function.

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