

Pseudotripling of optic disc: a case report

Pseudotriplicação do disco óptico: relato de caso

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ABSTRACT

Although coloboma of optic nerve head and/or chorioretinal tissue is not uncommon, duplication or pseudoduplication of the optic disc is rare. Here we report a rare case of pseudotripling of optic disc in a 50-year-old asymptomatic male patient. Orbital magnetic resonance image confirmed presence of only one optic nerve.

RESUMO

Embora o coloboma da cabeça do nervo óptico e/ou tecido coriorretiniano não sejam frequentes, a duplicação ou pseudoduplicação do disco óptico é rara. Relatamos aqui um caso raro de pseudotriplicação do disco óptico em um homem assintomático de 50 anos. A ressonância magnética orbital confirmou a presença de apenas um nervo óptico.

INTRODUCTION

Doubling of the optic disc is a rare clinical entity. Pseudodoubling of the optic disc is also relatively uncommon, and there are only a few reports of this finding. Pseudoduplication of the optic disc is considered to be generally associated with chorioretinal colobomas.⁽¹⁾ Differentiating between these two entities might face several challenges. Our case represents a unilateral pseudo-tripling of the optic disc. To our knowledge this is the first case of such a finding in the literature.

CASE REPORT

A 50-year-old male patient 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 has been wearing near glasses for several years. His past medical history was positive for newly-diagnosed type 2 diabetes mellitus. 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 the nasal macular region. No diabetic retinopathy was detected (Figure 1). In the fundoscopy of the left eye, the optic disc was relatively smaller than on 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 coursed 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.

We assure the patient this finding was not related to diabetes and would not impair visual function. After discussion with the patient, written informed consent was granted to use the fundus photography for publication purpose. The patient was scheduled to be examined for diabetic retinopathy after 1 year.

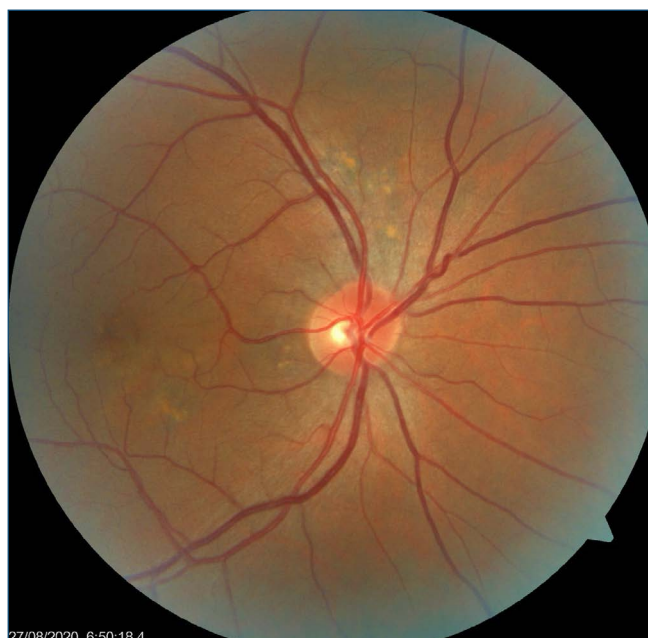


Figure 1. Fundus photography of the right eye



Figure 2. Fundus photography of the left eye.

DISCUSSION

Both duplication and pseudo-duplication of the optic disc are rare clinical entities. The lesions thought to be the “second” optic disc are usually located 0.5 to 3 disc diameters inferior to the true disc, show apparent cupping, and are associated with surrounding chorioretinal atrophy.⁽¹⁾ Typical clinical manifestations of pseudo-duplication of the optic disc include circumscribed, optic disc-like lesions. The vessels are usually radiating from the defect, and the central retinal vessels bridging the lesion.⁽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 described, in 1914, a case of doubling of the optic nerve disclosed at 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 pseudodoubling 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 computed tomography (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 nerve head 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 pseudo-doubling of optic disc. If in doubt, paraclinical 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 confirms the diagnosis. Perimetry can show double blind-spot, but this finding may be seen in the case of pseudodoubling.^(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 color Doppler imaging study can help 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⁾

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. To our knowledge, this is the first case of such finding described in the literature.

In conclusion, we presented a case of unilateral pseudotripling optic disc in an asymptomatic man. Our patient had no other ocular abnormality. True doubling of optic nerve head is a rare clinical condition. It can be differentiated from pseudodoubling by delicate funduscopy and/or paraclinical evaluation. Both conditions usually cause no harm to visual function.

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