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Optic disk drusen associated with congenital peripapillary staphyloma

Drusa de nervo ótico associada à estafiloma peripapilar congênito

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Abstract

We described a rare association between peripapillary staphyloma and optic disk drusen in a woman with 47 years old and normal vision.

Keywords: *Optic disk drusen; Scleral diseases; Retina; Ultrasonography; Congenital peripapillary staphyloma; Retinography; Autofluorescence*

Resumo

Nós descrevemos uma rara associação entre estafiloma peripapilar congênito e drusa de disco óptico em uma mulher de 47 anos de idade e visão normal.

Descritores: Drusas do disco óptico; Doenças da esclera; Retina; Ultrassonografia; Estafiloma peripapilar congênito; Retinografia; Autofluorescência

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Introduction

Optical disc drusen are acellular deposits located both intracellularly and extracellularly, first described by Muller in 1858.[1,2] Optical disk drusen occur in 3.4 to 24 per 1000 inhabitants, and are bilateral in approximately 75%.[3] Commonly asymptomatic, they may present with peripheral visual field deterioration and complications such as choroidal neovascularization, anterior ischemic optic neuropathy, retinal central artery occlusion, retinal central vein occlusion, subretinal neovascularization, and retinal hemorrhages.[3]

Optic nerve drusen have been described as occurring associated with ophthalmic artery aneurysm, hamartoma astrocytic, rotator atrophy, Birdshot chorioretinopathy, Cacchi-Ricci syndrome, congenital nocturnal blindness, familial macular dystrophy, glaucoma, nanophthalmos, central peripapillary serous retinopathy, pigmented retinochoroidal paravenosa atrophy, and thick cornea.[3] In the present study we will describe a case of optic nerve druse associated to peripapillary staphyloma (a condition in which an peripapillary ecleral thinning becomes ectasic.[4] The relationship between staphyloma and pathological myopia is widely described, with a 23% incidence of staphyloma in patients with pathological myopia[5], but this is the first description of optic nerve druse associated to staphyloma in a patient without pathological myopia.

Case Report

A 47-year-old white female, cleaning assistant, went to the ambulatory complaining of difficulty to read for two months. Visual acuity was bilateral (uncorrected) 20\20, there were no changes in both eyes to anterior biomicroscopy, intraocular pressure of 15 mmHg in the right eye and 14 mmHg in the left eye, retinal mapping in the right eye without changes, and mapping of retina of the left eye with the presence of optic nerve drusen and peripapillary staphyloma with no other changes (Figure 1). Fluorescein angiography (Figure 2), ocular ultrasonography (Figure 3) and visual campimetry 24.2 Humphfrey (Figure 4) were performed. Fluorescein angiography showed autofluorescent areas prior to intravenous injection of contrast, ultrasound demonstrated calcification, and campimetry showed increased blind spot, all of which were characteristic of this condition.[3]

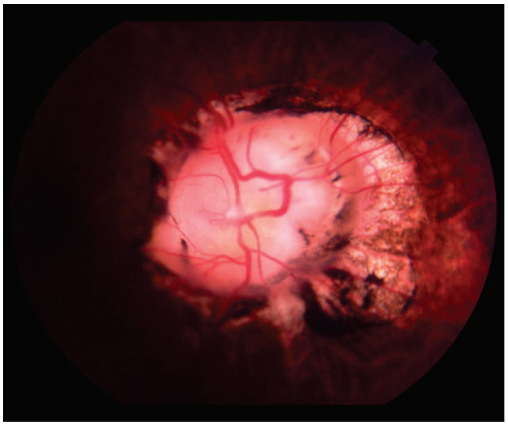

Figura 1: Peripapillary staphyloma and drusen of the optic nerve.

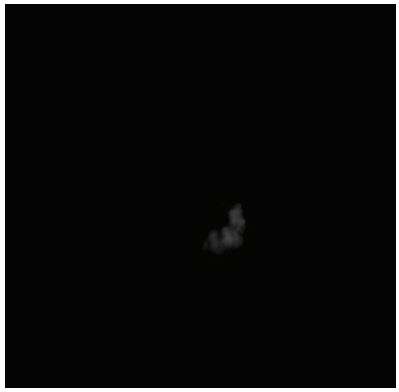

Figure 2: Autofluorescent areas characteristic of drusen of the optic nerve.

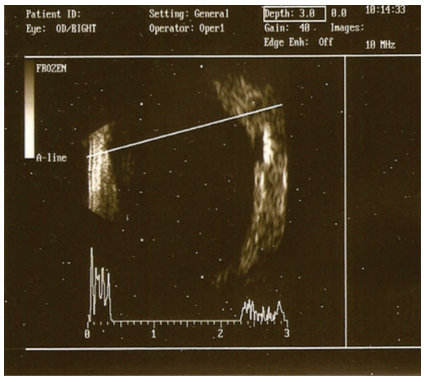

Figure 3: Presence of peripapillary staphyloma and hyperechoic lesion with acoustic shadow (white arrow) characteristic of optic nerve drusen.

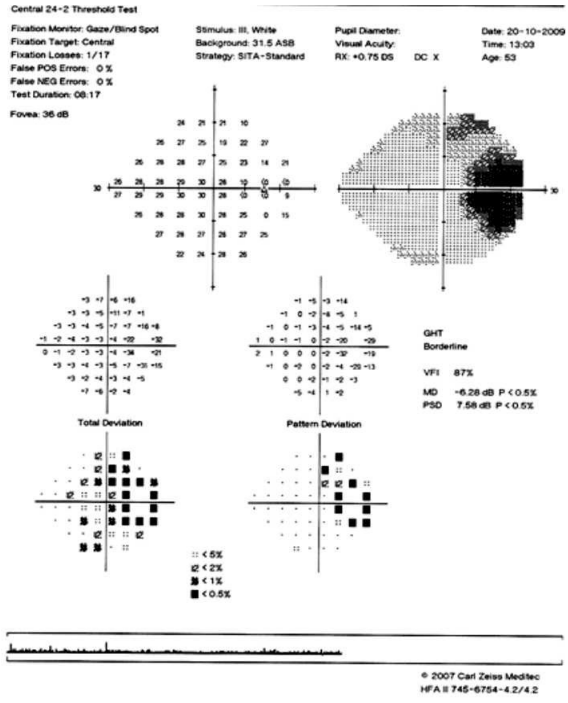

Figure 4: Visual field of the LE with increased blind spot characteristic of optic nerve druse.

Discussion

The peripapillary staphyloma belongs to the group of congenital anomalies of the papilla, and is extremely rare and differs from the papillary staphyloma of the high myopia, since the refraction found in the congenital ones is a mild myopia.[6]

To our knowledge, this is the first description of the association of peripapillary staphyloma and the druse of the optic nerve. In this case, the druse is unilateral, that is, only on the side affected by the staphyloma, so that the anatomical question seems to have been related to the appearance of the drusen.

The anatomical predisposition is one of the hypotheses for the pathogenesis of the drusen.[3] The first consideration would be the presence of a small scleral tunnel in the optic nerve head. Secondly, the vascular change in the papilla exit that would bring protein transudation, the vascular change was observed in our case.[3]

Another fact to be considered in the anatomical changes is the existence of contractile movements in the peripapillary staphyloma that may be related to the difference between the intraocular pressure and the CSF or contraction of the ocular muscles.[7]

In conclusion, this case demonstrates an extremely rare association, but with the development of imaging techniques the presence of optic nerve drusen in congenital peripapillary staphyloma may be more diagnosed.

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