



Pre- and juxtapapillary arterial loops in eyes with tilted disc syndrome and inferior staphyloma

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ABSTRACT

Purpose: To report and discuss the association between pre- or juxtapapillary arterial loops and tilted disc syndrome (TDS).

Observations: Three patients, aged 43–59 years, with both conditions were examined in a tertiary referral center, and underwent fluorescein angiography, optical coherence tomography (OCT) and/or OCT-angiography. They all presented with a typical inferior staphyloma associated with TDS and anomalies of insertion of retinal vessels. The vascular malformation consisted in one acquired arterial loop or cilioretinal collateral circulation occurring after central artery occlusion, and two more complex pre- and juxtapapillary arterial loops. In all cases, the vascular loops extended inferiorly, in the area of the staphyloma.

Conclusion and importance: We hypothesized that the local anatomical changes in the peripapillary area, observed in eyes with TDS and inferior staphyloma, could have promoted the occurrence and/or extent of the arterial loops.

1. Introduction

Tilted disc syndrome (TDS) was initially reported by Fuchs in 1882, in association with inferior staphyloma. Fuchs had noted that, in some eyes, a difference of 6 diopters could be observed between the inferior and superior parts of the fundus (reported in Apple DJ et al.).¹ Later, Curtin has classified the different types of staphylomas according to the part of the fundus affected by scleral ectasia. Inferior staphylomas corresponded to Type V of his classification, and were found in 2.6% of myopic eyes with staphyloma.² Numerous retinal complications have been reported in these eyes, including extensive pigmentary changes, choroidal neovascularization, macular serous retinal detachment, polypoidal choroidal vasculopathy, chorioretinal folds, and different vitreomacular interface changes.^{3–10} Changes in the shape of the eyeball that are associated tilted disc syndrome and inferior staphyloma can also be imaged by magnetic resonance imaging.^{11,12} Inferior staphylomas and tilted disc syndrome are frequently associated with changes in the papillary area, including inferior or infero-nasal conus and situs inversus.^{1,13} One of us has also reported the occurrence of a prepapillary arterial loop secondary to central retinal artery occlusion (CRAO) in a patient with tilted disc syndrome and inferior staphyloma.¹⁴ Herein, we

report two additional cases of arterial loop, TDS and inferior staphyloma and, based on these 3 cases, discuss the possible relationship between both conditions.

2. Case reports

Case 1 (summary, case previously reported in¹⁴).

A 49-year old woman was diagnosed with CRAO in a context of collagen vascular disease. The diagnosis was confirmed by fluorescein angiography. There was a typical TDS with inferior staphyloma. Two years later, she developed a prepapillary arterial loop or, more accurately, cilioretinal collateral circulation, extending in the inferior depigmented part of the fundus corresponding to the inferior staphyloma (Fig. 1).

Case 2. A 43-year old woman was referred for a peripapillary vascular loop diagnosed during a routine fundus examination. Tilted disc syndrome was present with inferior pallor of the fundus and abnormal insertion of the retinal vessels. Fluorescein angiography showed a juxtapapillary loop extending in the inferior part of the disc, in the area of the inferior staphyloma (Fig. 2). The patient was then lost to follow-up.

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Case 3. A 59-year old man was referred for optical coherence tomography (OCT) and fundus photography of a complex vascular anomaly that could be a pre- or juxtapapillary loop or a minor form of arteriovenous communication. The tilted disc syndrome was not obvious on fundus photography (Fig. 3), but was better seen on an SD-OCT oblique scan performed on the disc (Fig. 3 D), and an atypical insertion of the retinal vessels was also observed in the superior part of the disc. A typical inferior staphyloma extending towards the periphery was also observed on wide-field fundus photography. OCT-angiography (OCTA) allowed better analyzing the complex loops extending in the inferior part of the disc. Two years later, no change and no complication were noted.

3. Discussion

A prepapillary vascular loop may be acquired or, more frequently, congenital. The latter is part of congenital retinal vascular anomalies including, among others, macular macrovessels and congenital tortuosity.¹⁵ The loops are more often arterial than venous. They have recently been classified into 6 types according to their elevation, shape, number, location, type of vessel (arteriolar vs. venous), and presence of a vitreous traction. The 6 types have been described as flat, central, single, and isolated; flat, central, associated with retinal vasculature tortuosity; flat radial small; vitreous with a figure-of-eight configuration; vitreous with a corkscrew shape; or loop with vitreo-papillary traction, respectively.¹⁶ Different theories coexist for explaining the presence of loops, some considering them as part of the retinal vascular tortuosity syndrome, others as secondary to a deficient internal elastic lamina or to a vitreous traction. Congenital loops may occur as soon as the retinal vasculature develops, during the fourth month of pregnancy, i.e. the 100-nm embryonic stage.¹⁷ A histological report has supported the embryologic derivation of prepapillary loops from the retinal arterial system rather than from the hyaloid artery system.¹⁸

The present paper raised the question of a non-random occurrence of arterial loops or collaterals in eyes with congenital malformation of the disc. In acquired retinochoroidal collaterals with papillary retinal loops observed after central retinal vein occlusion, it was shown that nasal vortex veins served as extraocular exits of the drainage routes in all 10 involved eyes, with additional temporal vortex veins drainage in 3 eyes,¹⁹ suggesting a non-random occurrence. However, there is no study, to our knowledge suggesting that congenital malformation of the disc could influence the location of retinochoroidal collaterals in CRVO.

Tilted disc syndrome corresponds to an antero-posterior or infero-nasal tilting of the disc, but is usually also associated with other

anomalies of the disc or peripapillary area. First, an inferior or infero-nasal crescent (also called conus) is common. Second, anomalies of insertion of retinal vessels are also observed, the most common being situs inversus, corresponding to a congenital embryonic anomaly characterized by blood vessels initially emerging nasally from the optic disc before turning sharply temporally.²⁰ The relationship between these 2 conditions is not obvious. Indeed, tilted disc syndrome is considered a “forme fruste” of coloboma, i.e., a defect or a delay in the closure of the embryonic fissure, occurring during the sixth week of gestation and thus a few weeks before the development of the retinal vessels.¹ However, it is likely that changes in the morphology of the disc could modify the growth of retinal vascular buds leading to retinal vascular insertion anomalies. In case 1, a vascular arterial loop occurred secondary to CRAO, a phenomenon described as cilioretinal collateral circulation.²¹ However, we hypothesized that the preferential development of the loop towards the inferior staphyloma was not incidental in our case, but was promoted by anatomical conditions.

Areas of staphylomas correspond to a relative hypoplasia of the sclera, the choroid and the retina. The hypoplastic retina could be less resistant to the occurrence or extent of the loops. Cases 2 and 3 were probably congenital cases, because congenital loops are, by far, more common than acquired loops. However, they were diagnosed at the time of the first fundus examination of the patients, thus a congenital origin could not be assessed. Cases 1 and 2 presented with typical TDS while case 3 was less typical and showed anomalies of insertion of the retinal vessels and antero-posterior tilting that could be seen on SD-OCT optic disc cube analysis. The two potentially congenital cases could correspond to Type 3 loops, according to the classification suggested by Mansour and coworkers.¹⁶ But case 3 could also be classified as an arterio-arterial or arteriovenous communication.²² Indeed, differences between both conditions are not sharply delineated, and this case was not very different from a case described by Archer et al. as a group 2 arteriovenous communication in their widely accepted classification.²³

There is some confusion in the literature concerning vascular loops and their terminology. Indeed, terms as remodeled vessels, shunt vessels, collateral vessels are frequently used. The reasons may be to difficulty in differentiating small arteries and veins, the lack of longitudinal study allowing to analyze the acquired changes, and the poorly understood pathogenesis of the occurrence of these conditions. We decided to keep the term prepapillary arterial loops in the present report but remind fully aware that this term maybe not the most accurate.

All 3 cases did not show any vitreous traction, but showed inferior pallor of the fundus with a pattern of localized tessellated fundus associated with the inferior staphyloma. The association could be incidental.

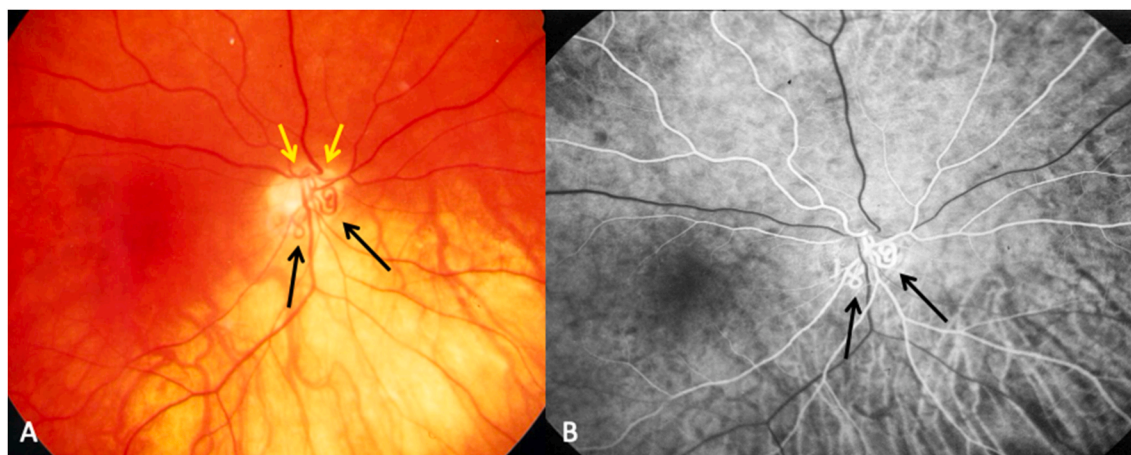


Fig. 1. Color fundus photography (A) and fluorescein angiography (B) of patient 1. The patient developed an acquired prepapillary loop (black arrows), 3 years after central retinal artery occlusion. A tilted disc was present (yellow arrows) with a typical inferior staphyloma. Reprint of Cohen SY. Acquired prepapillary arterial loop after central retinal artery obstruction. *Arch Ophthalmol* 1998; 116(10):1398-1399. Courtesy of JAMA. (For interpretation of the references to color in this figure legend, the reader is referred to the Web version of this article.)

However, it has been shown that tilted disc syndrome includes many changes in the morphology of the disc and juxtapapillary area, including a sloping of the lamina cribrosa posteriorly from the upper part to the lower part, a protrusion of the upper edge of the Bruch's membrane and choroid,²⁴ and a dissociation of optic disc margin components.²⁵ In other congenital disc malformations, anomalies of the small vessels have been recently reported. A case of hypoplasia of the optic nerve associated with a vascular loop and a fovea plana has been recently published, suggesting the presence of multiple developmental anomalies.²⁶ Moreover, in optic disc pits, OCT and OCTA show small, abnormal vessels both within and around the optic pits.²⁷ However, to our knowledge, there is no reported association between prepapillary arterial loops and other congenital disc anomalies. In the present study, we assumed that morphological changes of the disc, present in tilted disc syndrome, could have promoted the occurrence and/or the inferior extent of the arterial loop during embryogenesis or secondary to CRAO. However, additional cases are needed to fully confirm that vascular loops can be added to the numerous findings or complications associated with TDS and inferior

staphyloma.

4. Patient consent

Written informed consent was obtained from patients for publishing this case report and any accompanying images.

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Authorship

All authors attest that they meet the current ICMJE criteria for Authorship.



Fig. 2. Color (A), red-free fundus photography (B) and early and late frame of fluorescein angiography (FA, C, D) of patient 2. Tilted disc syndrome could be suspected based on the abnormal insertion of the superior retinal vessels (A). An inferior pallor corresponding to the inferior staphyloma was more obvious, better observed on red-free picture (B). FA showed that the vascular pre- and peripapillary loop was arterial (arrows, C), without involvement of retinal veins in the late frame of FA (D).

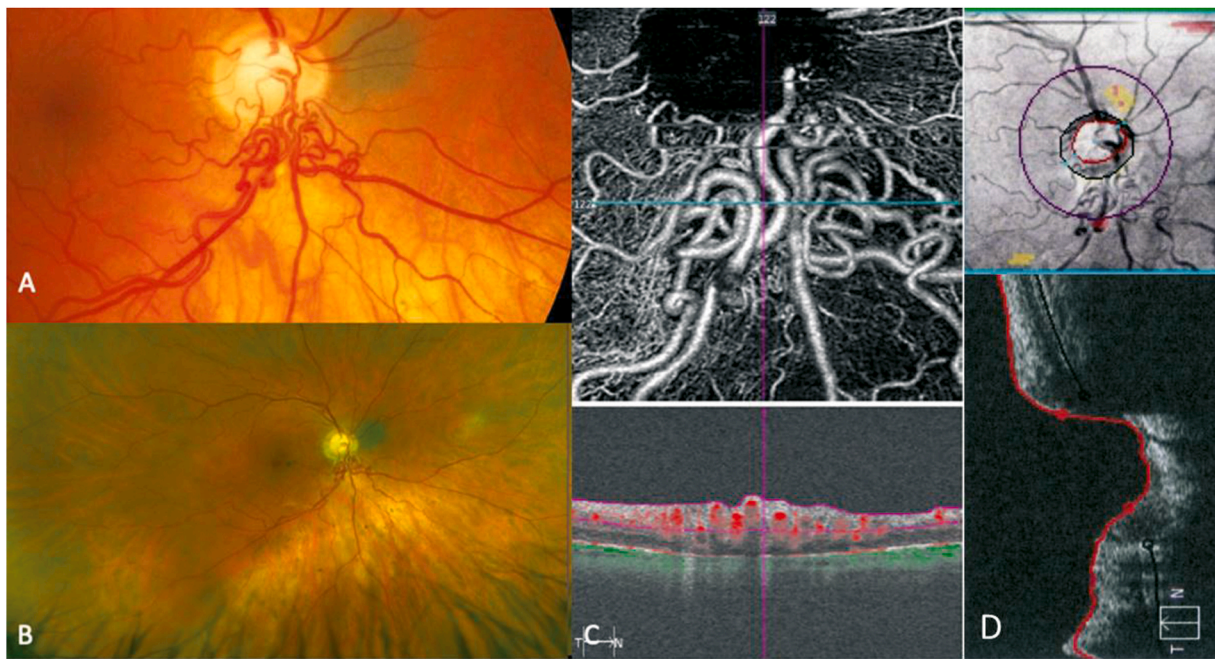


Fig. 3. Magnification of a color fundus photography (A), wide-field color photography (B), optical-coherence tomography-angiography (OCTA, C), and SD-OCT oblique scan passing through the disc (D) of patient 3. The tilted disc was not obvious on the 2-D pictures, but was visible on the spectral-domain OCT optic disc cube. An abnormal insertion of the superior vessels and an inferior staphyloma were visible. OCTA with a scan performed at the superior capillary network showed the complexity of the peripapillary loop that could also correspond to an arterio-arterial or arteriovenous communication. The oblique SD-OCT line better showed that the disc presents an anteroversion with the plan of the superior part of the disc appearing anterior to the plan of the inferior disc.

Declaration of competing interest

The following authors have no financial disclosures related to this study: SYC, SNB.

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