

PREPAPILLARY VASCULAR LOOP IN A GIRL WITH VENTRICULAR SEPTAL DEFECT – CASE REPORT

Isjanovski Igor^{1,2}, Pandilov Stefan¹

¹University Clinic for Eye Diseases, Skopje,

²Faculty of Medicine, Ss. Cyril and Methodius University in Skopje, Republic of North Macedonia

e-mail: dr.isjanovski@gmail.com

Abstract

A prepapillary vascular loop is a congenital variant of normal retinal vessels that is present on and/or around the optic disc. It is a rare vascular entity, with a prevalence of 1:2000 to 1:9000 eyes.

The aim of this paper was to describe a case of a 10-year-old girl in whom a unilateral prepapillary vascular loop was observed during a routine ophthalmological examination. Because of the anamnestic data of congenital heart anomaly, we performed a literature search of data available in several major medically relevant databases, but we did not come across another case where correlation of these two conditions was made. Considering that, this is the first case, known to us, where the following are present in the same patient: prepapillary vascular loop, eyelid ptosis and ventricular septal defect. A larger number of such patients is needed to draw a conclusion whether it is a systemic syndrome or a coincidental simultaneous presentation of multiple entities in the same patient.

Keywords: optic nerve, papilla, congenital abnormality, vascular loop

Introduction

Prepapillary vascular loops (PVLs) represent blood vessels with tortuous morphology that protrude into the vitreous in the area of the papilla. They can be either congenital, which are more common, and acquired^[1].

Acquired PVL usually occurs after occlusion of the central retinal artery or vein or their branches, optic nerve tumors, elevated intracranial pressure or cotton-wool ischemic changes of the retina of unknown origin^[1,2]. In these cases, the appearance of PVL is considered to be an attempt to reestablish a tissue reperfusion. Acquired PVLs are usually multiple, of smaller caliber and with pronounced tortuosity. Resorption hemorrhages or ghost vessels are often observed in the surrounding retina.

Congenital PVL is now considered a normal variant of the arrangement of retinal blood vessels in the papillary region. According to another classification, they belong to the group of congenital retinal vascular anomalies, together with: congenital tortuosity of blood vessels, arterio-arterial or arterio-venous bridging, macular macro-blood vessels and trifurcation of retinal vessels^[3] (Figure1).

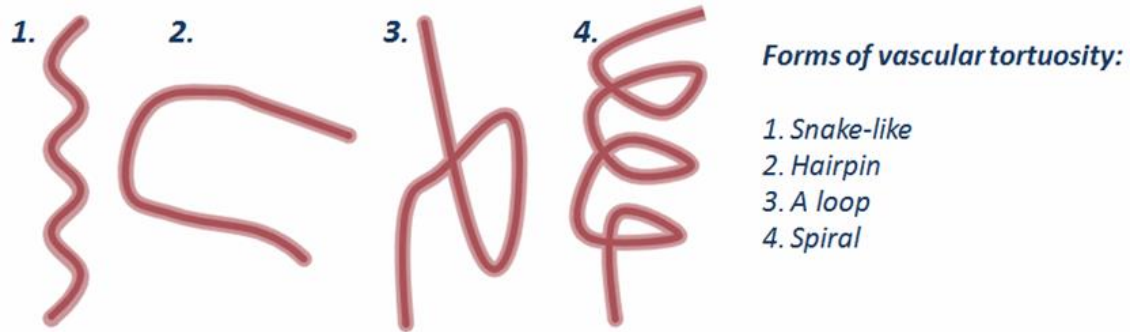


Fig. 1. Patterns of vascular tortuosity in prepapillary vascular loops

Morphologically, PVL can make up many patterns, which is why they have recently been classified into six different types^[4] (Figure 2).

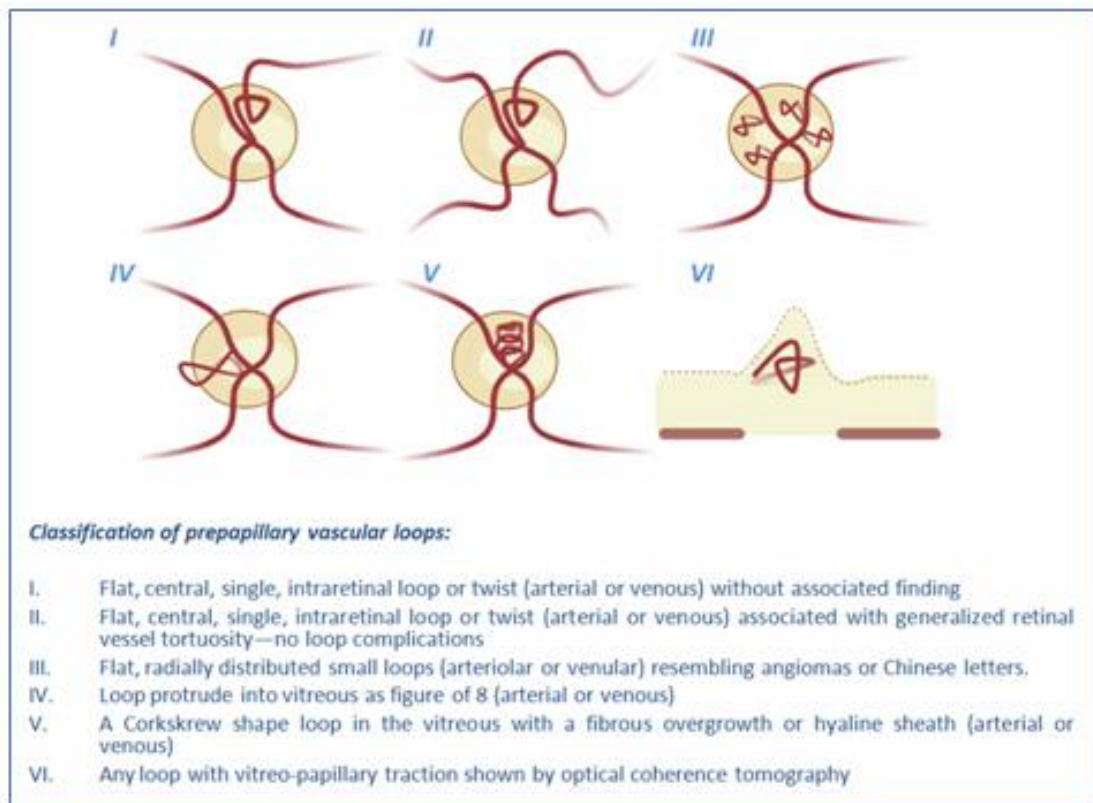


Fig. 2. New classification of prepapillary vascular loops according to Mansour *et al.* (Created and modified by: BioRender.com)

Case report

A 10-year-old girl came to the PHI University Clinic for Eye Diseases in Skopje for an ophthalmological examination. The anamnestic data obtained from the girl's parents pointed out the surgically corrected ventricular septal defect in infancy, for which she is regularly monitored by a pediatric cardiologist. During the ophthalmological evaluation, ptosis of the left upper eyelid was noted. Since it was present from an early age, it is probably a congenital ptosis. Best-corrected visual acuity was 6/6 bilaterally. Biomicroscopic examination of the anterior ocular segment and the motility of both eyeballs were within normal limits. A fundus examination of the left eye depicted a vascular prepapillary arterial

loop in the nasal sector of the papilla with prominence towards the vitreous body and a moderate mobility. The other blood vessels had an orderly course and lumen, without tortuosity; the macula was with preserved morphological characteristics and a cilioretinal artery present. Fundusoscopic findings of the right eye were unremarkable. A diagnosis of unilateral prepapillary arterial loop of size 1.9 mm type V was made. A native fundus photograph was taken of the patient to document the change (Figure 3). The condition was explained to the parents and advice was given for regular ophthalmological examinations.

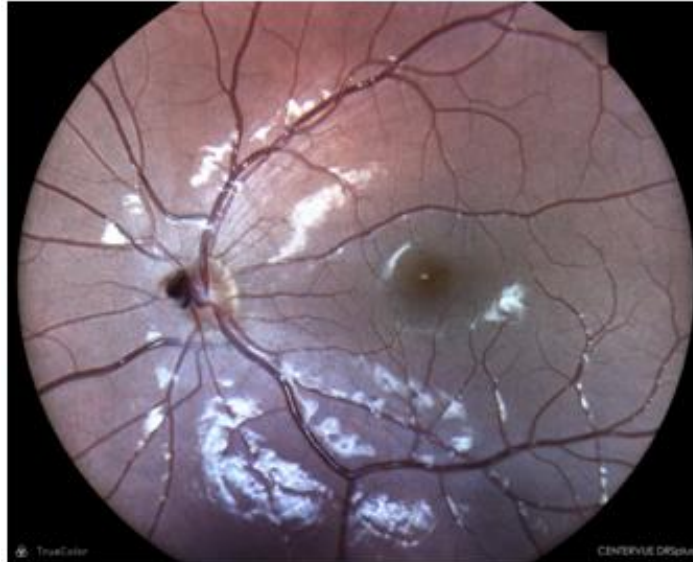


Fig. 3. A prepapillary arterial loop, 1.9 mm long, is observed near the optic nerve head, protruding towards the vitreous. Retinal blood vessels show normal diameter and flow, with a cilioretinal artery present. The macula exhibits regular morphology.

Discussion

PVL is a rather rare manifestation with a prevalence between 1:2000 to 1:9000 in the Western world^[5]. However, it should be noted that this entity is mostly asymptomatic, which is why it often goes undetected, and therefore it is considered to be much more prevalent. It is most often diagnosed accidentally during routine ophthalmological check-ups or when it consequently causes a certain retinal vascular event. It was first described in 1971 by Leibrich^[6]. At first, it was thought to be a variant and a remnant of the hyaloid blood vessels, but later with the help of fluorescein angiography and histopathological examination it was seen that it is a separate entity originating from the vascular network of the retina^[7].

According to the histo-anatomical study of Shakin *et al.* PVLs communicate with retinal arterial systems through small interconnects. The wall of the loops contains intima but lacks the internal elastic lamina, as is the case in the normal retinal artery. The perivascular connective tissue surrounding blood vessels in the eye typically contains lower levels of hyaluronic acid compared to the vitreous body. This connective tissue is continuous with the internal limiting membrane of the retina. In some individuals, this tissue can become more pronounced and form what is described as a "glial shell"^[8]. These histomorphological features support the hypothesis that PVLs originate from the retinal vasculature and are not remnants of the hyaloid vascular network.

From an embryonic point of view, Mann suggests that PVLs are created between the second half of the third and the first half of the fourth month of pregnancy. During this period the mesenchymal cells, as precursors of the endothelial cells of the retinal blood vessels,

inadvertently begin to grow forward into the supporting tissue of the Bergmeister papilla above the optic nerve head. In the absence of angiogenic factors, they then move back down to the disc and move towards the retinal tissue. Over time, Bergmeister papilla undergoes a process of involution and regression, leaving the PVL to persist in the Cloquet's canal^[9].

Over 90% of PVL are of arterial origin, while the rest are of venous origin. However, due to the dark discoloration and wider lumen, arterial PVLs might be misinterpreted as venous blood vessels^[10,11]. They can be unilateral or bilateral (less than 20%), one or more^[11]. Classically, arterial PVLs are between 1.5-7.88 mm in length, and venous ones are shorter, with a length of about 0.5 mm^[7]. Made up of an ascending branch that leaves a blood vessel of the optic disc, then forms a loop in the area of the vitreous cavity (mostly at the level of the Cloquet's canal) and a descending branch that returns to a blood vessel of the disc. The shape of the loop can be: a spiral or a screw, with the appearance of a figure eight, or a simple loop in the shape of a hairpin. In half of the cases, a small mobility of the loop is observed simultaneously with the heartbeat, while in 75% of patients with PVL the presence of a cilioretinal artery is also observed^[4,12]. Systemic association with other diseases is not routinely recorded. In our case, the presence of ventricular septal defect and ipsilateral congenital ptosis is probably an incidental finding rather than a systemic syndrome.

Regular follow-ups of patients with prepapillary vascular loops are necessary due to the possibility of complications such as occlusion of a branch of the central retinal artery, i.e. vein, hemorrhage in the vitreous body, formation of a macroaneurysm and its rupture, amaurosis fugax, appearance of hyphema, etc. Based on the up-to-date literature, even 10% of PVL patients develop some of the listed complications during their lifetime^[1,3,4,13,14]. Their occurrence is secondary to turbulent blood flow which in tortuous PVLs further leads to endothelial damage and tendency to thrombosis, thus fulfilling Virchow's triad for a thrombotic event. Therefore, when prepapillary hemorrhage in the vitreous or an occlusive vascular event near the papilla takes place, in the absence of other retinal pathology, PVL should be considered in the differential diagnosis as a possible cause.

Conclusion

PVL is a congenital variation of retinal blood vessels. It is mostly asymptomatic, which is why it is considered an underdiagnosed clinical entity. In this paper, we have described a case of unilateral prepapillary vascular loop in a girl with concomitant ptosis of the ipsilateral eyelid and previous surgery for ventricular septal defect. The presence of these three entities in this patient is probably coincidental, but to determine their potential syndromic association, further studies on a larger number of cases are needed.

Conflict of interest statement. None declared.

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