Neovascular glaucoma and the occurrence of twin vessels in congenital arteriovenous communications of the retina

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Abstract. Two cases with unilateral congenital arteriovenous communications of the retina are described. One of them developed a neovascular glaucoma. Both cases had twin vessels of the retina. The possible significance of twin vessels in these cases is discussed.

Abbreviations: AVC - Arterio-Venous Communication, AVCR - Arterio-Venous Communication of the Retina

Introduction

Congenital arteriovenous communications of the retina (AVCR's) are rather rare. Up till now about a hundred cases have been described in literature. Most cases remain stable during life but in some spontaneous alterations have been mentioned [1, 2]. Congenital AVCR's should be separated from AVCR's of secondary origin, e.g. after venous occlusions, if only because these secondary AVCR's have more spontaneous changes.

Recently the term twin vessels has been coined in ophthalmology [3]. A twin vessel is one pair of vessels consisting of a retinal arteriole and venule that run an adjacent course for more than one disc diameter and that are separated by less than the diameter of one venule (Fig. 1). Because of vessel crowding near the optic disc a twin vessel should by definition be more than two disc diameters away from the optic disc [3]. In the human body this vessels pattern is quite common, especially in connective tissue, whereas in the eye it is only customary in the conjunctiva but not in the retina [4].

The purpose of this paper is to report an up to my knowledge not earlier described case of neovascular glaucoma as a complication of a congenital AVCR as well as to point out the existence and possible relevance of twin vessels in two cases with a congenital AVCR.

Fig. 1. Fluorescein angiogram of one (between arrows) of the three twin vessels in the left eye of case 2. Note partially filled venule parallel to arteriole for trajectory longer than one disc diameter and with less space between the two vessels than the diameter of the venule.

Case reports

Case 1

At age 8 this boy was referred in 1973 because he was found to have a subnormal visual acuity in his right eye during a routine examination by his school doctor. The visual acuity was R eye 3/60 with Sph + 3.25, L eye 0.9 with Sph + 2.25. In his right eye a grade III AVCR [1] was found (Fig. 2) with spontaneous pulsations of strongly dilated veins. Five years later he came back because of black specks in his right visual field sometimes accompanied by a stinging sensation. The visual acuity in his right eye was 0.1, no rubeosis was seen and the intraocular pressure was 10 mm Hg in his right and 12 mm Hg in his left eye. In the right fundus extensive intraretinal haemorrhages were noted together with some previously undescribed white sheathing along some AVCR vessels. At the end of 1978, four months later, he came back to the outpatient department because of mixed pericorneal redness, right pupillary dilatation, pain and nausea and vomitus. He was found to have in his right eye only light perception, rubeosis of the iris, uveal

Fig. 2. (a) Fluorescein angiogram of AVCR on top of the optic disc in right eye of case 1 in 1973.

Fig. 2. (b) Fluorescein angiogram of AVCR in macular area in right eye of case 1 in 1973.

Fig. 3. Fluorescein angiogram of right eye of case 1 in 1987 after neovascular glaucoma and spontaneous involution of AVCR.

ectropion and an intraocular pressure of 44 mm Hg. The fundus looked the same as before. Diathermal coagulation of the ciliary body under a scleral flap was performed at the 12 o'clock position without much pressure lowering effect. Within a month cryocoagulation was applied to the ciliary body in the lower two quadrants. In the following ten months the rubeosis disappeared and the tortuous vessels gradually became thinner. At the end of 1979 the right visual acuity amounted to hand movements, the pressure was 22 mm Hg and the AVCR vessels were described as partly obliterated with a silver wire aspect. In the next few years the intraocular pressure gradually went up till 50 mm Hg without subjective complaints.

In 1987 there was no light perception any more in his right eye and the rubeosis was gone. The pressure was 18 mm Hg and in the fundus cupping of the disc with small shunt vessels as well as involutionary gliosis of the AVCR was visible (Fig. 3). Also a twin vessel was present in the same fundus above the temporal arcade (Fig. 4). The left eye showed no abnormalities.

Case 2

A 24 year old man visited the outpatient department in 1986 because of intermittent redness and a "heavy" feeling at night in his right eye. Three years earlier the redness started with a frequency of once a week when he was

Fig. 4. Twin vessels in same eye as Fig. 3.

tired but the intervals became progressively shorter. It was constantly present since three months. In 1977 he had been hit with a fist on the same eye after which the general practitioner kept him in bed for three weeks. The ophthalmologist who saw him afterwards noted dilated vessels on his right optic disc that he considered to be a congenital anomaly. The patient was healthy and had had no epileptic insults. An uncle of his had a cerebral haemorrhage at age three which resulted in a spastic left arm and leg.

On examination his visual acuity was R eye 1.0 with Sph + 0.75, L eye 1.25 E. The right conjunctiva showed in the nasal inferior part dilated vessels. The right pupil was a bit wider than the left one; the reactions on light were normal and there was no afferent defect. On the right disc many tortuous vessels were seen (Fig. 5) resembling a grade III AVCR [1]. The left fundus was normal but for three twin vessels (Fig. 1). Neurological examination including CT scans showed no abnormalities.

Discussion

Two grading systems for the severity of AVCR's have been proposed [1, 2]. Mansour et al. call an anastomosis between a small arteriole and venule grade I; grade II an anastomosis between a branch artery and a branch vein;

Fig. 5. AVCR in right eye of case 2.

grade III a diffuse marked dilatation of the whole vascular system. In the classification of Archer et al. group 1 had capillaries between the arterial and venous side, group 2 had direct arteriovenous communications while group 3 showed severe malformations leading to loss of visual acuity [2]. Group 3 thus seems more or less to coincide with grade III.

AVCR's grade III have more visual loss and more systemic AVC's than those of grade I and II [1]. Both our cases had an AVCR grade III but no systemic lesions and one kept a good vision in the affected eye.

In literature glaucoma has been mentioned in conjunction with congenital AVCR only once without further description [1]. Venous occlusion, however, seems to be present in 6% of all hitherto described cases of AVCR [1]. Thus on theoretical grounds neovascular glaucoma could have been expected as a complication of congenital AVCR.

As the extent of the haemorrhages was not noted, it was difficult to judge from the old description in the file of case 1 if there was a picture of a full-blown central vein occlusion before the neovascular glaucoma started. Also it was not clear if one had specifically looked for retinal new vessels. However, the course of this neovascular glaucoma is rather uncommon. Case 1 received only diathermal and cryo treatment of the ciliary body. No attempts at coagulation of the retinal periphery were made and probably only the neovascular membrane in the angle was partly treated during the cyclocoagulations. This could explain the disappearance of the rubeosis but

not so well that the eye became quiet again without vitreous haemorrhages or other sequelae. One could assume a continuing process of capillary closure resulting in progressive retinal ischaemia and a gradual disappearance of the still hypothetical vasoproliferative factor. Capillary non-perfusion in the retinal periphery in cases of AVCR has been mentioned once [10].

Spontaneous closure of a congenital AVCR has been described before [1, 5, 6, 7, 8, 9, 10]. In most cases an involutionary sclerosis of the AVCR vessels was noted attributable to the extraordinary haemodynamic relations that have been demonstrated [2]. Haemorrhages along the dilated vessels were mentioned [1, 2, 5, 7] as well as in an area peripheral to the AVCR [8]. In two cases a central retinal vein occlusion has been reported [10, 11] once with extensive neovascularization on the disc and the retina. In the latter case panretinal Xenon lamp photocoagulation was performed after which a vitreous haemorrhage occurred [10]. The intraocular pressure was not mentioned.

It is difficult to judge the significance of twin vessels in these two cases of congenital AVCR's. We do not know exactly the prevalence of twin vessels in the (ophthalmic) population. Actually a twin vessel was found in two eyes of two out of 36 subjects with non-retinal disorders (cataract, squint) who attended an outpatient ophthalmological clinic [3]. This would point to a prevalence of 5.5% twin vessels for the ophthalmic patients or to 2.8% for their eyes. When the rarity of congenital AVCR is taken into account it is in any case remarkable that both our patients had twin vessels.

Twin vessels are considered to be congenital anomalously running vessels without demonstrable pathology. On fluorescein angiography these vessels showed a normal calibre in comparison with vessels in the neighbourhood, normal circulation times, no leakage of dye and no staining in the late period. It is noteworthy that both von Hippel-Lindau disease and congenital AVCR are disorders of a vascular system that develops during the 15th week of gestation from the same primitive retinal mesenchymal cells as the retinal and thus also the twin vessels. I also have seen a patient with a Sturge Weber syndrome with a twin vessel in one eye. One might speculate that twin vessels could be in some subjects a aspecific marker for a congenital retinal vascular disorder. Further studies will have to clarify this. Arteriovenous communications in their turn, both in the retina as elsewhere, have also been mentioned in connection with hereditary haemorrhagic teleangiectasia or Rendu-Osler-Weber syndrome [1].

The congenital AVCR might in some cases be called a twin vessel because these vessels sometimes have a parallel course for quite some distance. I think they should be excluded as they are a separate entity with clearly abnormal vessels.

Congenital AVCR seems to be a sporadic disorder that is bilateral only in 4% of all cases [1]. In connection with systemic AVC's a dominant inheritance has been claimed but without substantiation with exact data [12]. Our cases were unilateral. In case 1 the twin vessels were in the same eye as the AVCR but in case 2 in the contralateral eye. If this has a significance remains to be demonstrated in the future.

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