Nettleship collaterals: circumpapillary cilioretinal anastomoses after occlusion of the central retinal artery

Nicola K Ragge, William F Hoyt

Abstract

In extremely rare cases, after occlusion of the central retinal artery, a complete ring of peripapillary anastomotic channels develops. One such case is described and a proposed term given to these channels - 'Nettleship collaterals' - after the man who first described them. These collaterals are dilated pathways within the terminal capillary networks of the posterior ciliary arteries and the branches of the central retinal artery. Formation of the channels is believed to be promoted by prelaminar obstruction of the central retinal artery.

A century ago at St Thomas' Hospital, London, Nettleship¹ illustrated the ophthalmoscopic appearance of circumpapillary arterial collaterals in a patient with an occluded central retinal artery. He described these collaterals as occurring between the 'sclerochoroidal ring' and branches of the central retinal artery.

Fourteen years later in Lausanne, Gonin² published a similar drawing. The branches of the central retinal artery were narrowed proximally on the optic disc and then after a short retinal course widened abruptly to almost normal calibre. A succession of 'petites anses ou de crochets' (small hooks) could be seen emerging from the margin of the disc and joining the main arterial branches at their points of enlargement. Indeed some of the arterial branches were fed by a confluence of several collaterals.

Although limited peripapillary arterial collaterals occasionally form after occlusion of the central retinal artery or one of the main branches on the disc,³⁻⁹ documentation of com-

plete rings of these collaterals is rare. Since Nettleship and Gonin these exceptional cases have appeared in the form of sketches,¹⁰ colour photographs,^{11 12} a description with no illustration,¹³ and fluorescein angiograms.^{12 14}

A new case of circumpapillary cilioretinal arterial collaterals surrounding an anteriorly obstructed central retinal artery is discussed and recorded photographically. The term 'Nettleship collaterals' is proposed for these anastomotic channels.

Case history

An 85-year-old woman was examined 8 months after an occlusion of her right central retinal artery, which left her with no light perception in her right eye. Her general health was otherwise good and she did not have hypertension, diabetes, or valvular heart disease. She was taking aspirin, 150 mg daily and prednisolone, 5 mg daily. Her visual acuity was 20/30 (corrected) in the left eye.

Her right optic disc (Figs 1 and 2) was pale and atrophic. It was surrounded by a distinctive corona of multiple arterial-to-arterial collaterals. These collateral vessels arose from just within the disc margin and via a tortuous route joined and supplied the branches of the retinal artery in the peripapillary zone. In some instances the collaterals appeared to take a spiral course (for example, the vessel at 1 o'clock) or were coiled initially and then straightened just before joining the retinal arterial branch (for example, the vessel at 11 o'clock). One collateral (at 10 o'clock) arose from inside the disc margin in a

Figure 1 Photograph of the right fundus of the patient 8 months after occlusion of the central retinal artery, showing 'Nettleship collaterals'.

Figure 2 Diagram of the right fundus of the patient to illustrate with further clarity the 'Nettleship collaterals'. Arrows indicate the presumed direction of blood flow within them.

Neuro-ophthalmology Unit, Departments of Neurosurgery and Ophthalmology, School of Medicine, University of California, San Francisco, CA 94143 USA

N K Ragge
W F Hoyt

Correspondence to: William F Hoyt, MD, Department of Neurological Surgery, c/o The Editorial Office, 1360 Ninth Avenue, Suite 210, San Francisco, CA 94122, USA.

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similar way to the others, traversed the disc margin, and then looped sharply back on itself to anastomose with a small direct branch from the central retinal artery. In general the retinal arterial branches were attenuated and had significant irregularity of calibre; this irregularity was especially prominent in the inferior branches. Where the branches were joined by the collaterals some attained a wider calibre (for example, those at 12 o'clock and 1 o'clock). The central retinal artery appeared on the disc as a ghost vessel, and the initial segments of its main branches contained only a narrow column of blood: most of their lumens appeared to be occluded. Fluorescein angiography showed that filling of both retinal and choroidal circulation was markedly delayed: dye first appeared 28 seconds after the injection. Both circulations were filled simultaneously.

In the left fundus there was evidence of occlusion of a superior branch of the retinal artery as well as age-related macular degeneration. The results of investigations for temporal arteritis and heart disease were negative but carotid doppler studies revealed 50% stenosis of the right carotid artery and haemodynamically insignificant disease of the left carotid artery.

Discussion

'Nettleship collaterals' are multisectorial cilioretinal collateral vessels that arise from posterior ciliary vessels in the optic disc, course over the edge of the disc, and join the branches of the retinal artery in the peripapillary area. These collaterals form, after occlusion of the central retinal artery, by enlargement of selected channels within the capillary networks already existing in the optic disc and in the peripapillary superficial nerve fibre layer. The networks are supplied by the posterior ciliary arteries and the proximal trunks of the retinal artery.^{14 15} Although there appears to be significant flow in these vessels Nettleship collaterals usually represent an ineffective attempt to revascularise the retina via the posterior ciliary circulation after occlusion of the main stem of the central retinal artery. Presumably the attempt at revascularisation fails for two reasons. First, the amount of blood flow is insufficient; second, it reaches the retina after the death of the retinal neurons. In most reported cases (Table 1) the eyes remain totally blind unless a cilioretinal artery is present. One patient with well-maintained visual

acuity, described by Marmor and colleagues,¹² is an exception to the preceding generalisations.

Data from experiments on cats¹⁶ and rhesus monkeys¹⁷ and from observation of patients^{7 9} suggest that collaterals probably develop within days or weeks after occlusion of the central retinal artery, although in most reported cases they are only detected months later. Presumably the vessels become more conspicuous in the later stages because of their increasing width and their increasing contrast to an optic disc that is becoming atrophic.

Formation of collaterals around an obstructed central retinal artery is promoted by several factors: complete obstruction of the vessel;¹⁷ a suitable pressure gradient in the alternative blood supply, here the branches of the posterior ciliary artery;¹⁶ and possibly local vasoproliferative factors stimulated by ischaemia in the peripapillary retina.⁷ Complete obstruction of the central retinal artery can be caused by different types of emboli or by thrombosis. Calcific emboli (making up 9% of all emboli) have been traditionally associated with more severe and permanent visual loss and with the formation of collaterals.⁷ In fact valvular heart disease, the most frequent source of calcific emboli, was present in many of the reported patients with Nettleship collaterals.^{2 10 11} Cholesterol emboli can also be the cause of sectorial peripapillary collaterals.⁷ In this patient atheromatous disease of the carotid artery may have been the source of the embolus. The cilioretinal collaterals described by Hayreh¹⁴ were seen in a 14-year-old girl whose central retinal artery was occluded as a result of herpes zoster ophthalmicus. Presumably the mechanism of occlusion in that patient was arteritis combined with *in situ* thrombosis.

What then are the special circumstances that promote the formation of Nettleship collaterals? There are two possible causes: a rare anterior site of occlusion or a rare anatomical variation.

In the first situation, as suggested by Nettleship¹ in his original description, the central retinal artery is obstructed at an unusually anterior site. Obstruction of the central retinal artery most commonly occurs behind the lamina cribrosa.^{18 19} The central retinal artery then refills, either by recanalisation or via presumed anastomotic vessels^{14 15 17 20-25} from the posterior ciliary circulation, sometimes indirectly via pial branches. These anastomoses form anterior to the obstruction but too deep to be visible on the surface of the disc. However if the obstruction is anterior to the lamina cribrosa the anastomoses are unable to feed into this section of the central retinal artery. This forces the formation of alternative cilioretinal collaterals from the rim of the disc. The anterior occlusion is visible on the disc in this case and in other cases of Nettleship collaterals.^{1 14} Further, an anterior embolus was visible in all of the cases of sectorial collateral formation after retinal artery occlusion described by Arruga and Sanders⁷ and in some additional reported cases.^{4 10}

The second possible reason for the formation of Nettleship collaterals depends on the wide variations in branches of the central retinal artery and posterior ciliary arteries.²⁶ It may be that in rare individuals no potential exists for communi-

Table 1 Summary of clinical details for reported patients with Nettleship collaterals after occlusion of the central retinal artery

Author (year)	Age of patient (sex)	Valvular heart disease	Carotid artery disease	Hypertension	Visual acuity after CRAO	Time from occlusion to first detection of collateral formation
Nettleship (1891) ¹	? (M)	?	?	?	?	12 months
Gonin (1905) ²	64 (F)	+	-	-	NLP	3 months
Harms (1914) ¹³ (case 3)	47 (F)	+	-	-	HM†	11 months
Jensen (1938) ¹⁰ (case 2)	61 (M)	+	-	+	CF, then 6/12*	24 months
Larsen (1969) ¹¹	40 (M)	-	-	+	NLP	3 months
Hayreh (1969) ¹⁴	14 (F)	-	-	-	?	9 months
Marmor <i>et al</i> (1985) ¹²						
Case 1	69 (M)	-	+	-	6/6	?
Case 2	67 (F)	-	-	+	6/60	?
This case (1991)	85 (F)	-	+	-	NLP	8 months

*Cilioretinal artery present. †No illustration supplied. CRAO=central retinal artery occlusion, NLP=no light perception, HM=hand movements, CF=count fingers.

cation between these two vascular networks except in the peripapillary area. In such cases communication would open up in this area after obstruction of the central retinal artery regardless of the site of occlusion.

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