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Branch Retinal Vein Occlusion in a Japanese Patient with Neurofibromatosis 1

Fumihiko Mori, Motofumi Kawai, Eiichi Sato,
Sho Igarishi, Taiichi Hikichi and Akitoshi Yoshida

Department of Ophthalmology, Asahikawa Medical College, Asahikawa, Japan

Background: To report an Asian patient with branch retinal vein occlusion secondary to neurofibromatosis 1.

Case: A 64-year-old woman presented with a loss of vision in her right eye of 9-month duration. A diagnosis of neurofibromatosis 1 was made. A general medical examination showed no abnormalities except the signs of neurofibromatosis 1.

Observations: Fundus examination of the right eye revealed irregularities of the venous caliber, partial sheathing of the corresponding vein and macular edema. Multiple dilated and tortuous collateral channels and an arteriovenous communication bridged the perfused and nonperfused retina. Fundus examination of the left eye showed a tortuous vein in the temporal region of the fovea. Fluorescein angiography of the right eye confirmed delayed filling in a superotemporal artery and in the corresponding vein. The temporal region of the fovea had large areas of capillary loss. The collaterals were tortuous and mimicked a neovascularization. Fluorescein angiography of the left eye confirmed that the vein in the temporal part of the fovea was tortuous and not leaking. A diagnosis of branch retinal vein occlusion of the superotemporal vein was made.

Conclusions: Neurofibromatosis 1 should be considered in the differential diagnosis of retinal vascular occlusive disease without other risk factors. **Jpn J Ophthalmol 2001;45:634–635** © 2001 Japanese Ophthalmological Society

Key Words: Branch retinal vein occlusion, neurofibromatosis 1, retinal vascular occlusive disorder.

Introduction

Retinal and choroidal abnormalities have been reported in patients with neurofibromatosis 1 worldwide.[1–3] Systemic vascular ischemic manifestations of neurofibromatosis have been reported in the aortic, cerebral, and renal vasculature.[4] Two cases of retinal vascular occlusive disorders have been reported in patients with neurofibromatosis.[5,6] This is the first report of a Japanese patient with branch retinal vein occlusion and neurofibromatosis 1.

Case Report

A 64-year-old woman presented with a loss of vision in her right eye of 9-month duration. The best-corrected visual acuity was 20/200 OD and 20/20 OS. A diagnosis of neurofibromatosis 1 was made on the basis of multiple cafe au lait spots, plexiform neurofibroma, and Sakurai-Lisch nodules of the iris in both eyes. Fundus examination of the right eye revealed irregularities of the venous caliber, partial sheathing of the corresponding vein and macular edema. Multiple dilated and tortuous collateral channels were present, and an arteriovenous communicating artery bridged the perfused and nonperfused retina. Fundus examination of the left eye showed a tortuous vein in the temporal part of the fovea. Fluorescein angiography (FA) of the right eye confirmed delayed filling in a superotemporal artery

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Correspondence and reprint requests to: Fumihiko MORI, MD, PhD, Department of Ophthalmology, Asahikawa Medical College, Midorigaoka Higashi 2-1-1-1, Asahikawa-shi 078-8510, Hokkaido, Japan

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and in the corresponding vein. The temporal part of the fovea had large areas of capillary loss. The collaterals were tortuous and mimicked a neovascularization (Figure 1A). A diagnosis of branch retinal vein occlusion of the superotemporal retinal vein was made. FA of the left eye confirmed that the vein in the temporal part of the fovea was tortuous and not leaking (Figure 1B).

A general medical examination showed no abnormalities (eg, hypertension, diabetes, or hyperlipemia) except for the signs of neurofibromatosis l. The results of magnetic resonance imaging of the brain and orbits were unremarkable.

Discussion

We have presented the findings in a case of retinal vascular occlusive disorder secondary to neurofibromatosis 1. The retinal manifestations associated with neurofibromatosis include retinal astrocytic hamartoma, combined hamartoma of the retina and retinal pigment epithelium, and retinal capillary hemangiomas.[1,2] Only two cases of retinal vascular occlusive disorders have been reported in patients with neurofibromatosis 1.[5,6] To our knowledge, this is the first report of an Asian patient with neurofibromatosis and retinal vascular occlusive disorder.

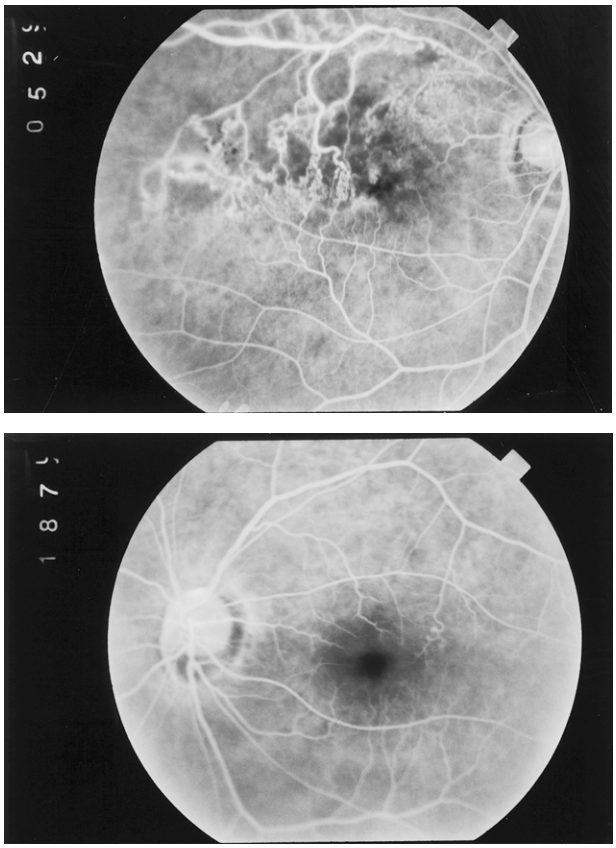

Figure 1. (**Top**) Fluorescein angiogram of right eye showing temporal fovea with large areas of capillary loss and tortuous collaterals mimicking neovascularization. (**Bottom**) In left eye, vein in temporal part of fovea is tortuous and not leaking.

Systemic vascular involvement of the aortic, cerebral, and renal vasculature has been reported in patients with neurofibromatosis.[4] In our patient, a branch retinal vein occlusion in the superotemporal vein of the right eye and a tortuous vein in the temporal part of the fovea of the left eye were found. These pathologic features in the fundus of the eye are similar to the vascular abnormalities in other organs (eg, vascular occlusive disease, capillary closure, compensatory arteriovenous communications). The retinal vascular occlusion in this patient may be similar to that causing ischemia in the kidneys, brain, heart, and large vessels in patients with neurofibromatosis 1. The growth of Schwann cells in the vascular lesions results in a vascular abnormality in neurofibromatosis 1.[4] The glial proliferation may induce occlusion of the retinal vessels as is the case with other vessel tissues in neurofibromatosis 1.

In our patient, the development of the retinal vascular occlusion may have been the response to a branch vein occlusion. A general medical examination did not show any additional risk factors for vaso-occlusive disease except for the signs of neurofibromatosis 1. Whatever the mechanism of the vascular occlusive lesions in neurofibromatosis, we propose that more patients with neurofibromatosis 1 should be examined for retinal vascular occlusive disorders. Furthermore, neurofibromatosis 1 should be considered in the differential diagnosis of retinal vascular occlusive disease without other risk factors.

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