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Frosted Branch Angiitis Associated with Harada Disease-like Manifestations Recurs 10 Years Later

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Purpose: To describe a case with frosted branch angiitis occurring in association with manifestations of Harada disease.

Case: A 41-year-old man developed marked iridocyclitis, frosted branch angiitis and retinal white dots, together with serous detachment of the sensory retina in the posterior pole of both eyes.

Observations: Fluorescein angiography demonstrated multiple retinal pigment epithelial leakage points in the early phase followed by late dye pooling in the serous detachment, consistent with the manifestations of Harada disease. The patient experienced recurrence of the same combination of manifestations 10 years later. On both occasions, he showed mononuclear cell increase and protein elevation in the cerebrospinal fluid on spinal tap.

Conclusion: Fluorescein angiography and spinal tap supported the diagnosis of Harada disease in the patient. The present patient could be diagnosed as having either Harada disease with frosted branch-like response or frosted branch angiitis with serous retinal detachment as occurs in Harada disease. **Jpn J Ophthalmol 2002;46:682–684** © 2000 Japanese Ophthalmological Society

Key Words: Frosted branch angiitis, Harada disease, recurrence.

Introduction

Frosted branch angiitis is a clinical entity of uveitis with unknown cause. The characteristic sign is diffuse perivenous sheathing of the whole retina, usually involving both eyes, seen often in children and also in adults.[1] It has been proposed that it would be more accurate to call this condition "acute frosted retinal periphlebitis,"[2] but the original term, "frosted branch angiitis," is still in wider use. A frosted branch-like appearance is also observed as a clinical sign in association with viral infections, autoimmune diseases, or malignancy such as lymphoma.[3] In contrast with such secondary frosted branch angiitis or frosted branch-like appearance or response, the initial clinical entity occurring in otherwise healthy young patients should be called "acute idiopathic frosted branch angiitis."[3] We report a patient who experienced a 10-year recurrence of acute idiopathic frosted branch angiitis in association with serous retinal detachment as observed in Harada disease.

Case Report

A 41-year-old man noticed metamorphopsia in both eyes in February 1989. The best-corrected visual acuity was 20/30 in the right eye and 20/70 in the left eye. The intraocular pressure was 18 mm Hg in both eyes. Both eyes showed many fine keratic precipitates and aqueous cells as well as Koeppe iris nodules. Serous detachment of the sensory retina around the optic disc was associated with retinal perivenous sheathing and retinal yellowish spots (Figures 1A, 1B). Fluorescein angiography disclosed multiple retinal pigment epithelial leakage points in

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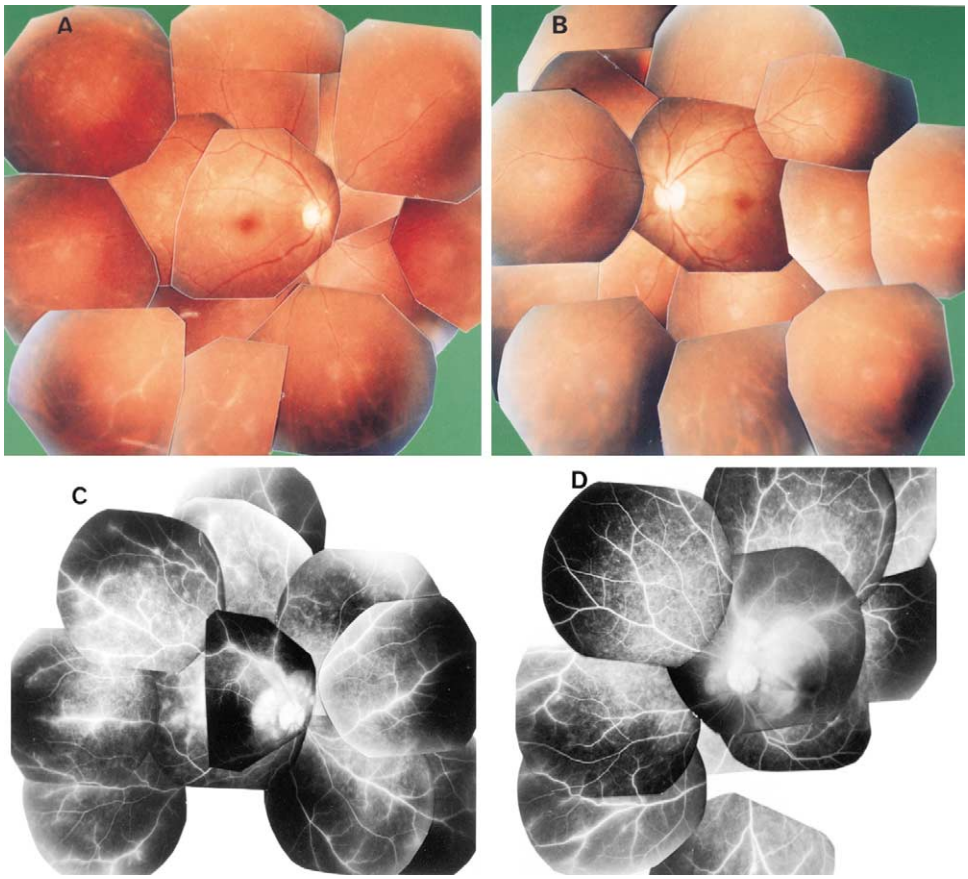

Figure 1. February 1989. Fundus photographs of the right (**A**) and left (**B**) eyes of a 41-year-old man show serous retinal detachment around the optic disc and frosted branch angiitis mainly involving the midperiphery. Fluorescein angiography of the right (**C**) and left (**D**) eyes demonstrates retinal pigment epithelial damage with dye pooling and perivenous staining.

the early phase followed by late dye pooling in the retinal detachment, typical of Harada disease (Figures 1C, 1D). In addition, perivenous staining and spotty hyperfluorescence were observed to correspond to the perivenous sheathing and retinal spots, respectively. Results of systemic tests showed mononuclear cell increase and protein elevation in the cerebrospinal fluid on spinal tap, and moderate bilateral sensory hearing loss for high pitch sounds. Results of chest x-ray and laboratory examinations were normal. He had no lymphadenopathy or skin lesions. Human leukocyte antigen (HLA) typing showed A2, A24(9), Bw46, DRw11(5), and DRw8. With intravenous prednisolone tapered from 200 mg daily, the serous retinal detachment and perivenous sheathing resolved, and the visual acuity returned to 20/20 in both eyes. Depigmented spots appeared in the midperipheral fundus after the subsidence of inflammation.

He was in good health for the next 10 years until December 1999, when he noticed blurred vision in both eyes. The best-corrected visual acuity was 20/30 in the right eye and finger counting in the left eye. The intraocular pressure was 15 mm Hg in both eyes. He showed many fine keratic precipitates and aqueous cells in both eyes, with lower half posterior iris synechiae in the right eye, serous detachment of the sensory retina in the posterior pole of the left eye, and perivenous sheathing with many retinal white spots in the midperiphery of both eyes (Figures 2A, 2B). Spinal tap disclosed mononuclear cell increase and protein elevation. He was given intravenous prednisolone tapered from 200 mg daily, and recovered the visual acuity of 20/20 in both eyes.

Discussion

The patient showed the combination of clinical signs typical of acute idiopathic frosted branch angiitis, such as no systemic manifestations, acute onset, bilateral involvement, iridocyclitis, diffuse perivenous

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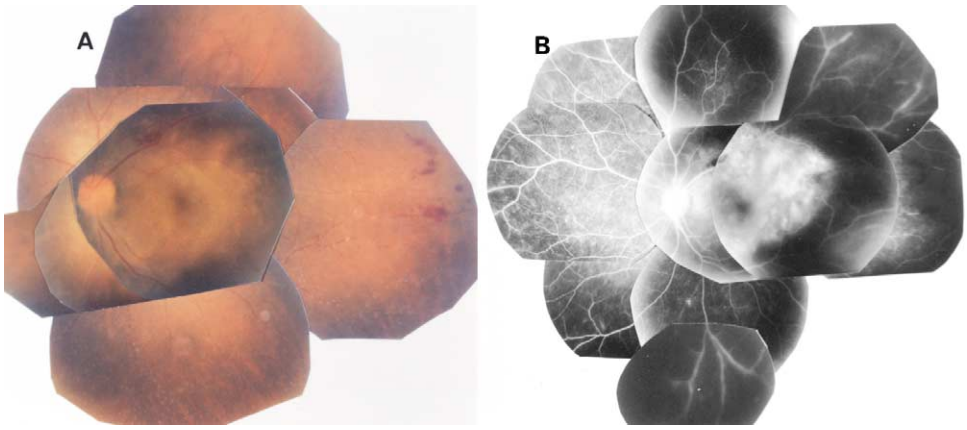

Figure 2. December 1999. In the same patient, fundus photographs (**A**) in the left eye show perivenous sheathing and hemorrhage as well as serous retinal detachment in the posterior pole. Fluorescein angiography (**B**) shows retinal pigment epithelial damage with dye pooling and perivenous staining.

sheathing, and good response to corticosteroids. In addition, he presented serous retinal detachment with fluorescein angiographic features typical of Harada disease. Serous retinal detachment has been found in 10 of 58 patients (17%) with acute idiopathic frosted branch angiitis reported previously.[1] The present patient did not have any systemic signs to support the diagnosis of other diseases, such as sarcoidosis or malignant lymphoma, which sometimes show frosted branch-like appearance.

Harada disease could be diagnosed in this patient based on fluorescein angiography and spinal tap both at the initial presentation and at the time of the recurrence. Furthermore, at the initial presentation, he showed sensory hearing disturbance for high-pitch sounds and developed depigmented spots in the midperipheral fundus after the inflammation subsided. These features are all characteristic of Harada disease. The patient, however, had no typical skin lesions and was therefore diagnosed as having incomplete Harada disease based on recent criteria.[4] In addition, the absence of HLA-DR4 in this patient is rather unusual for Harada disease.

Harada disease recurs usually as iridocyclitis. Its late recurrence as serous retinal detachment is uncommon but has been reported.[5] However, as in the present patient, it is unusual for Harada disease to recur as serous retinal detachment after an interval as long as 10 years. He developed depigmented spots in the midperipheral fundus but no apparent depigmentation of the whole fundus. In Harada disease, early treatment with a high dose of corticosteroids does indeed result in less depigmentation of the fundus.

The patient, described in the initial report as having acute idiopathic frosted branch angiitis, also developed depigmented areas of the fundus after the subsidence of the inflammation.[1] Such depigmentation is a frequent sequela to Harada disease. Serous retinal detachment as seen in Harada disease sometimes has been described as a clinical sign in acute idiopathic frosted branch angiitis.[1] These previous cases and the present patient would support the clinical linkage between acute idiopathic frosted branch angiitis and Harada disease. Both diseases might be located at either end of the spectrum of a common disease. The present patient may thus be diagnosed as having either Harada disease with frosted branch-like response or acute idiopathic frosted branch angiitis with serous retinal detachment as seen in Harada disease.

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