Birdshot Retinochoroidopathy
—A Possible Relationship to Ocular Sarcoidosis—

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Summary: This report concerns four patients who exhibited characteristic ocular signs of birdshot retinochoroidopathy and were diagnosed as suffering from sarcoidosis on the basis of histological evaluation of lymph node biopsy. The findings in the present series suggest that sarcoidosis may be one of the cause of birdshot retinochoroidopathy. Two patients exhibited reddish appearance of the posterior fundus ("sunset-glow" fundus). This report suggests when "sunset-glow" fundus is seen in the patient with uveitis, the presence of sarcoidosis should be considered besides that of Harada's disease and sympathetic ophthal mia.

Key words: birdshot retinochoroidopathy—sarcoidosis—uveitis—sunset glow fundus—ocular sarcoidosis

Introduction

Sarcoidosis is a systemic granulomatous disease in which ocular involvement is common. There are many reports of ocular sarcoidosis (Franceschetti and Babel, 1949; Laval, 1952; Chumbley and Kearns 1972; Brownstein and Jannotta, 1974; Letocha et al. 1975; Sanders and Shilling, 1976; Gass and Olson, 1976; Marcus et al. 1982). Sarcoidosis may affect the eye in many ways. They may be anterior segment involvement which usually takes the form of a granulomatous uveitis. Another common sign of sarcoidosis is the Busacca nodule of the iris. Finally, the features of posterior segment involvement include chorioretinitis, periphlebitis, chorioretinal granuloma, chorioretinitis “en taches de bougie”, macular edema, papilledema, optic nerve granuloma, vitreous cellular infiltration, and rarely, retinal neovascularization.

In 1981, Spalton and Sanders reported that birdshot retinochoroidopathy can be found in some patients exhibiting histologically confirmed sarcoidosis. This report concerns four patients suffering from sarcoidosis who showed the characteristic ocular signs of birdshot retinochoroidopathy. The purpose of this report is to discuss a possible causal relationship between birdshot retinochoroidopathy, “sunset-glow” fundus and ocular sarcoidosis.

Materials and Methods

All patients were women. At the time of the examination, the patients ranged in age from 58 to 72 years. The period of follow-up ranged from 11 to 70 months.
(with a mean of 38 months). Visual acuity test, slit-lamp examination, fundus photography and fluorescein fundus angiography were performed in all patients. Farnsworth Panel D-15 examination, adaptometry, stereo fundus photography, electroretinograms and electrooculograms were performed in three patients, while Goldmann visual field examination was carried out in 2 patients. Laboratory studies, chest X ray, transbronchial lung biopsy, Ga scintigram and lymph node biopsies were carried out in all patients in the Department of Internal Medicine of the Kurume University Hospital. Sarcoidosis was histologically confirmed in all patients by lymph node biopsy. All examinations were performed with the informed consent of the patients. Two patients were treated with injectable steroid prior to the present study (Cases 3, 4). Three patients were treated with oral steroid under the control of the physician (Cases 1, 3 and 4).

Case Report

Case 1. In April 1982, we examined a 72-year-old woman with a history of decreased vision of the right eye and floaters of both eyes for two months. She has had gastric ulcer twelve years ago. Her family history was not significant. On examination, her best corrected visual acuity was 0.3 for the right eye and 0.3 for the left eye. Her right eye deviated laterosuperiorly, she did not complain of diplopia. On slit-lamp examination, many fine and medium-sized keratic precipitates were found in both eyes. Marked cellular reactions were present in both anterior chambers. In right eye, she presented with exfoliated material at the latero-inferior portion of the dilated pupil. Both lenses were clear. Cells were present diffusely in both vitreous cavities. On gonioscopy, there were small localized peripheral anterior synechias (PAS) in both eyes. There were no nodules and no signs of iris neovascularizations. In the posterior pole, many flat, yellow-white patches of the retinal pigment epithelium and choroid were present bilaterally (Fig. 1A and B). She exhibited bilaterally macular edema. Her both posterior fundi were reddish in color. Fluorescein fundus angiography showed no abnormality in the early retinal phase. In the late phase, there were hyperfluorescences corresponding to areas involved ophthalmoscopically (Fig. 2A-C). On Farnsworth Panel D-15 test, she showed tritan in both eyes. Electroretinogram showed reduced a and b wave amplitude in the right eye. Her electrooculogram values were 1.9 and 1.48 for the right and left eye, respectively. Goldmann visual field examination and adaptometry revealed no abnormalities in either eye. On laboratory studies, angiotensin converting enzyme activity (ACE) was 110 Iu/1 (normal 8.3-21.4 Iu/1). Her chest X-ray showed no hilar lymphadenopathy. On Ga scintigram, the accumulation was present in both eyes, both parotid glands and the lung. Biopsy of subcutaneous lymph node in the buttock disclosed epithelioid granuloma with Langhans type giant cells suggestive of sarcoidosis.

She was treated with prednisolone, 60 mg daily. In March 1987, she visited this Department for follow-up study. Her best corrected visual acuity was 0.3 and 0.7 for the right and left eye, respectively. There were no cellular reactions in either anterior chamber. Senile cataract appeared in both lenses. The number of cells in the vitreous cavity decreased in both eyes. Yellow-white patches continued being present on ophthalmoscopic examination, but the cystoid macular edema disappeared bilaterally. The reddish appearance of the posterior fundus ("sunset-glow" fundus) did not change. Fluorescein fundus angiography showed hyperfluorescence corresponding to the yellow-white patches in the late retinal phase. The patient presented with tritan in both eyes on Farns-
Fig. 1. Case 1. A, right eye  B, left eye. Both fundus photographs showing many depigmented areas and reddish appearance in the posterior pole.

Fig. 2. Fluorescein fundus angiogram of the right eye in Case 1. Angiogram showing no abnormal fluorescence in retinal arteriovenous phase (A). Angiogram showing many round hyperfluorescence corresponding to the depigmented areas in the venous phase (B). Angiogram showing many round hyperfluorescence and cystoid macular edema (arrow) around the temporal half of the fovea in late phase (C).
worth Panel D-15 test. Electroretinogram showed reduced a and b wave amplitude in the right eye. Electrooculogram values were 1.58 and 1.39 for the right and left eye, respectively.

Case 2. In August 1981, a 58-year-old woman stated on interview that she suffered for one month of floaters in both eyes. The past history revealed pulmonary tuberculosis at 25 years of age and rheumatoid arthritis at 55 years of age. She suffered ocular injury of the right eye at 15 years of age. She was treated in the past for gastric ulcer and dysautonomia. Her family history was not significant. On examination, her best corrected visual acuity was 0.2 and 1.0 for the right and left eye, respectively. She presented nubeculae at the center of the cornea. There were cells in both anterior chamber, keratic precipitate was not present. Both lenses were clear. Cells were present diffusely in both vitreous cavities. Nodules, PAS, or neovascularization were not found in either eye on gonioscopy. Ophthalmoscopically, flat, yellow-white patches were present at the posterior pole and temporal equatorial fundus of the right eye; in the left eye, patches were present at the posterior pole and nasal equatorial fundus. Disc and macula were not involved on either side. Fluorescein angiography showed no abnormality in the early retinal phase, but revealed hyperfluorescence corresponding to patches seen ophthalmoscopically in the late phase. Farnsworth Panel D-15 test, Goldmann visual field examination and adaptometry revealed no abnormalities. Electroretinogram showed reduced a and b wave amplitude and disappearance of the oscillatory potential in the left eye. Electrooculogram values were 1.8 and 1.9 for the right and left eye, respectively. In laboratory studies, ACE was 32IU/l. The chest X-ray disclosed no hilar lymphadenopathy. Diagnosis of sarcoidosis was made on the basis of the evaluation of the supraclavicular lymph node biopsy.

She was not treated with steroid. In June 1987, her best corrected visual acuity was 0.5 and 0.9 for the right and left eye, respectively. Cells were present diffusely in both vitreous cavities. On fundus examination, yellow-white patches in both eyes were still presented.

Case 3. In October 1985, we examined a 60-year-old woman with a history floaters of both eyes for the past six months. Her past history and family history were not significant. On examination, her best corrected visual acuity was 1.0 and 1.0 for the right and left eye, respectively. There were the medium-sized keratic precipitates in both eyes. A few cells were present in both anterior chambers. On gonioscopy, she had some nodules in the right eye. Both lenses were clear. In both vitreous cavities, cells were diffusely present. There were snow ball opacities in the right eye. Ophthalmoscopically, many flat yellow-white patches were present in both eyes at the inferior equatorial fundus. The patient exhibited papilledema bilaterally. There were findings of retinal periphlebitis in both eyes. Fluorescein fundus angiography showed no abnormality in the early retinal phase, but there was hyperfluorescence corresponding to the yellow-white patches and leakage from the disc in both eyes in the late phase. On laboratory studies, ACE was 21.4 IU/l. Chest X-ray did not reveal hilar lymphadenopathy.

Diagnosis of sarcoidosis was made on the basis of the evaluation of supraclavicular lymph node biopsy. The patient was treated with prednisolone, 30 mg/every other day. In October 1986, her best corrected visual acuity was 1.0 for both the right and left eye. Cells disappeared from both anterior chambers. Cells were diffusely present in both vitreous cavities. Snow-ball opacities and iris nodule in the right eye did not change. On fundus examination, the papilledema remained pre-
### TABLE 1
**Summary of cases**

<table>
<thead>
<tr>
<th>Case</th>
<th>Age</th>
<th>Sex</th>
<th>Visual Acuity</th>
<th>Vitreous Cavity</th>
<th>Pigment Epithelium and Choroid</th>
<th>Retina</th>
<th>Optic Nerve</th>
<th>ERG</th>
<th>B.O.G</th>
<th>Final Visual Acuity</th>
<th>Period of Follow up</th>
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<tbody>
<tr>
<td>1</td>
<td>72</td>
<td>F</td>
<td>0.3</td>
<td>0.3</td>
<td>cells</td>
<td>Depigmented areas in the posterior pole</td>
<td>Cystoid macular edema</td>
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<td>58</td>
<td>F</td>
<td>0.2</td>
<td>1.0</td>
<td>cells</td>
<td>Depigmented areas in the posterior pole and equatorial fundus</td>
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<td>1.0</td>
<td>cells</td>
<td>Depigmented areas in the equatorial fundus</td>
<td>papilledema</td>
<td>1.0</td>
<td>1.0</td>
<td>12</td>
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<tr>
<td>4</td>
<td>69</td>
<td>F</td>
<td>0.7</td>
<td>0.4</td>
<td>cells</td>
<td>R.E.: Depigmented areas in the posterior pole</td>
<td>Cystoid macular edema</td>
<td>1.36</td>
<td>1.23</td>
<td>0.9</td>
<td>0.3</td>
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### TABLE 2
**Characteristic signs of the sarcoidosis**

<table>
<thead>
<tr>
<th>Case</th>
<th>Mutton fat precipitate</th>
<th>Nodule of the iris</th>
<th>Tentorial PAS</th>
<th>Snow ball opacities</th>
<th>Periphebitis</th>
<th>Retinal neovascularization</th>
<th>ACE (IU/l)</th>
<th>B.H.L</th>
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<td>+</td>
<td>29.4</td>
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B.H.L.: Bilateral hilar lymphadenopathy
sent in both eyes and yellow-white patches were not changed. On the other hand, retinal periphlebitis disappeared from both eyes.

Case 4. In August 1986, a 69-year-old woman presented with a 3 month complain of decreased vision and floaters of the left eye. She suffered from essential hypertension for a period of ten years and has been taking medicine for hypertension for the last five years. Her sister suffered from diabetes mellitus, otherwise, her family history was not significant.

On examination, her best corrected visual acuity was 0.7 and 0.4, for the right and left eye, respectively. She exhibited many fine keratic precipitates on both eyes, there was no cellular reaction in either anterior chamber. Upon pupillary dilation, she presented with exfoliated material at the temporal margin of the right eye. Both lenses were clear. There was a slight cell reaction in the right vitreous cavity. In the left vitreous cavity, cells were diffusely present and there were snow ball opacities. On gonioscopy, she had the tentorial PAS in the left eye, but there was no nodule and no neovascularization in either eyes. On fundus examination, flat yellow-white patches were present at the posterior pole in the right eye; in the left eye, same patches were scattered at the temporal equatorial fundus and the retinal neovascularization was seen along the equatorial superotemporal retinal vein. There was cystoid macular edema in the left eye. Her both posterior fundi were reddish in color ("sunset-glow" fundus). Fluorescein fundus angiography showed hyperfluorescence corresponding to the yellow-white patches in the late phase in both eyes, and severe leakage from the retinal neovascularization in the left eye. On Farnsworth Panel D-15 test, she showed tritan in the left eye. In laboratory studies, ACE was 29.4 Iu/l. Her chest X-ray showed hilar lymph adenopathy.

She was diagnosed as suffering from sarcoidosis on the basis of the evaluation of the biopsy of the supraclavicular lymph node, and she was treated with prednisolone, 40 mg daily. On July 1987, her best corrected visual acuity was 0.9 and 0.3 for the right and left eye, respectively. There was no cellular reaction in either anterior chamber. Vitreous cells and the fundus appearance were unchanged.

The summary of the findings and the characteristic signs of sarcoidosis for these four cases are shown in Tables 1 and 2.

Discussion

The cause of birdshot retinochoroidopathy is unknown and sarcoidosis has been differentiated from birdshot retinochoroidopathy as a cause of the latter in the past reports (Kaplan and Aaberg, 1980; Ryan and Maumenee, 1980; Gass, 1981). However, the four cases of the present series which presented with characteristic ocular signs of birdshot retinochoroidopathy were diagnosed as sarcoidosis histologically.

Ophthalmologist often diagnoses sarcoidosis by the presence of the characteristic ocular signs, such as mutton fat precipitates, Busacca nodule of the iris, tentorial PAS in the chamber angle, snow ball opacities in the vitreous cavities, and retinal periphlebitis. In the present series, however, two cases (Case 3 and 4) presented with the ocular signs which are characteristic for sarcoidosis, while the other two cases did not exhibited the signs in question. This suggests that the diagnosis of sarcoidosis should not be excluded on the basis of the absence of the characteristic ocular signs of sarcoidosis. It is difficult to ascertain systemic signs of sarcoidosis from the ocular signs alone, ACE, while X-ray findings are often unreliable. In fact, without the availability of the
lymph node biopsy, sarcoidosis would not be diagnosed for Case 2. Finally, the findings in the present series suggest that sarcoidosis may be one of the causes of birdshot retinochoroidopathy.

Yellow-white patches of the retinal pigment epithelium and choroid did not respond in our series to the steroid and did not show the hyperfluorescence in the early retinal phase of fluorescein fundus angiography. It may be suggested therefore that yellow-white patches are not constituted by the granuloma formation in the choroid.

We found “sunset-glow” fundus in the posterior fundus of two patients (Case 1 and 4). To the best of our knowledge, the reddish appearance of the posterior fundus (“sunset-glow” fundus) accompanied with sarcoidosis or birdshot retinochoroidopathy was never described before. We found that the degree of “sunset-glow” fundus did not relate to the number of the yellow-white patches and electrooculogram values. We feel that the cause of this funduscopic feature may not be the diffuse disturbance of the retinal pigment epithelium. Although we could not find a definite cause for sunset-glow fundus in the present series, it is important to note that when sunset-glow fundus is seen in patients exhibiting uveitis, the presence of sarcoidosis should be considered besides that of Vogt-Koyanagi-Harada disease and sympathetic ophthalmia.

References


