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Cancer-associated Nummular Loss of the Retinal Pigment Epithelium

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Purpose: To report a case of cancer-associated nummular loss of the retinal pigment epithelium.
Methods: A 47-year-old man with a history of hepatocellular carcinoma presented with three weeks of bilateral visual loss. His best-corrected visual acuity was 20/40 in each eye. He had multiple round confluent grayish-brown patches at the level of retinal pigment epithelium, and no pigmented choroidal lesions. Fluorescein angiography showed circular areas of transmission defect and indocyanine green angiography showed early hyperfluorescence, corresponding with the multiple round confluent patches.
Conclusions: We report a case of visual paraneoplastic syndrome which showed nummular loss of the pigment epithelial cells which distinguishes the clinical component of BDUMP syndrome.
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Key Words: Bilateral diffuse uveal melanocytic proliferation, Cancer-associated retinopathy, Paraneoplastic syndrome

Bilateral diffuse uveal melanocytic prolifelation (BDUMP) is a bizarre paraneoplastic ocular syndrome occurring in patients with systemic carcinoma.[1-5] Gass et al.[2] identified five cardinal ocular signs that accompany visual loss in BDUMP: (1) multiple, round or oval, subtle, red patches at the level of the retinal pigment epithelium in the posterior fundus; (2) a striking pattern of multifocal areas of early hyperfluorescence corresponding with these patchs; (3) development of multiple, slightly elevated, pigmented and nonpigmented uveal melanocytic tumors, as well as evidence of diffuse thickening of the uvea; (4) exudative retinal detachment; and (5) rapid progression of cataract. Pathologic studies show proliferation of benign melanocytes in the outer choroid that is histopathologically unrelated to the primary nonocular carcinoma. We report a case with features of BDUMP, where the predominant finding was round patches of retinal pigment epithelium loss.

Case Report

47-year-old man presented with a complaint of bilateral visual loss for the previous three weeks. He had undergone a liver transplantation to treat hepatocellular carcinoma 1 year ago. His best-corrected visual acuity was 20/40 in each eye.

Ocular motility, anterior segment slit-lamp examination findings, and intraocular pressure were normal. He had multiple round confluent grayish-brown patches at the level of retinal pigment epithelium, and there was serous detachment in the posterior pole. No pigmented choroidal lesions were found. Fluorescein angiography showed circular areas of transmission defect; staining of fine deposits within these regions without significant leakage was also noted. Interspersed between these areas were garlands of preserved retinal pigment epithelium (Fig. 1).

Electroretinography revealed a bilateral decrease of a- and b-waves under both photophic and scotopic conditions. Goldmann perimetry revealed concentric constriction in both eyes. Optical coherence tomography showed zones of retinal pigment epithelium loss alternating with areas of thickened retinal pigment epithelium and deposit of debris that corresponded with the areas of transmission defect seen during fluorescein angiography (Fig. 1). Two months later, the patient's visual acuity decreased to 20/80 OD and 20/100 OS. Fundus photograph, Fluorescein angiography and Indocyanine green angiography showed that both the number and the size of multiple round patches increased (Fig. 2). The patient received an injection of subtenon triamcinolone acetonide (40 mg). Within 4 months from the onset of visual symptoms, the patient died from hepatic failure. No autopsy was done.

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Discussion

Visual paraneoplastic syndrome associated with cancer is classified by susceptible retinal cells. Cancer-associated retinopathy (CAR) affects photoreceptors. Melanoma-associated retinopathy (MAR) is thought to affect bipolar cell function, and bilateral diffuse uveal melanocytic proliferation (BDUMP) targets the uveal tract.[6,7] Each disease shows different clinical findings and progresses depending on the presence of susceptible retinal cells.[6,7]

Gass et al.[2] described five specific ocular features associated with BDUMP. But not all cases show such characteristics. Gass et al.[2] considered two fundus findings to be important: (1) multiple, round or oval, subtle, red patches at the level of retinal pigment epithelium in the posterior fundus; (2) a striking pattern of multifocal areas of early hyperfluorescence corresponding with these patches. In this case, only two such fundus findings were observed.

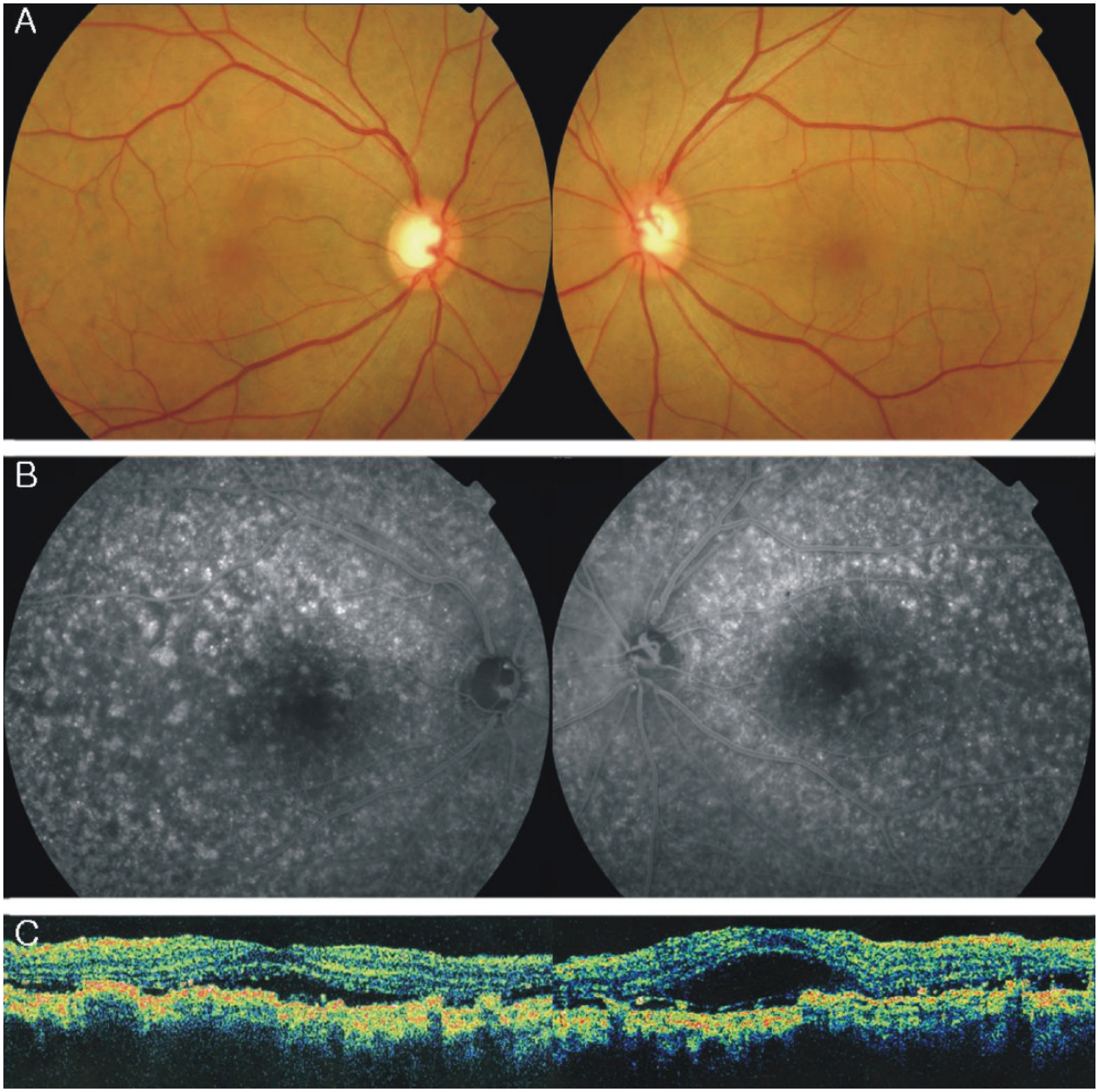

Fig. 1. Initial visit. (A) Fundus photograph. Multiple round confluent grayish-brown patches are noted at the level of retinal pigment epithelium. There were no pigmented choroidal lesions. (B) Fluorescein angiography. Hyperfluorescence due to window defects associated with the widespread retinal pigment epithelium damage is seen corresponding to the lesion. No significant leakage. (C) Optical coherence tomography. OCT image showed zones of retinal pigment epithelium loss alternating with areas of thickened retinal pigment epithelium and deposit of debris that correspond with the multiple round confluent patches.

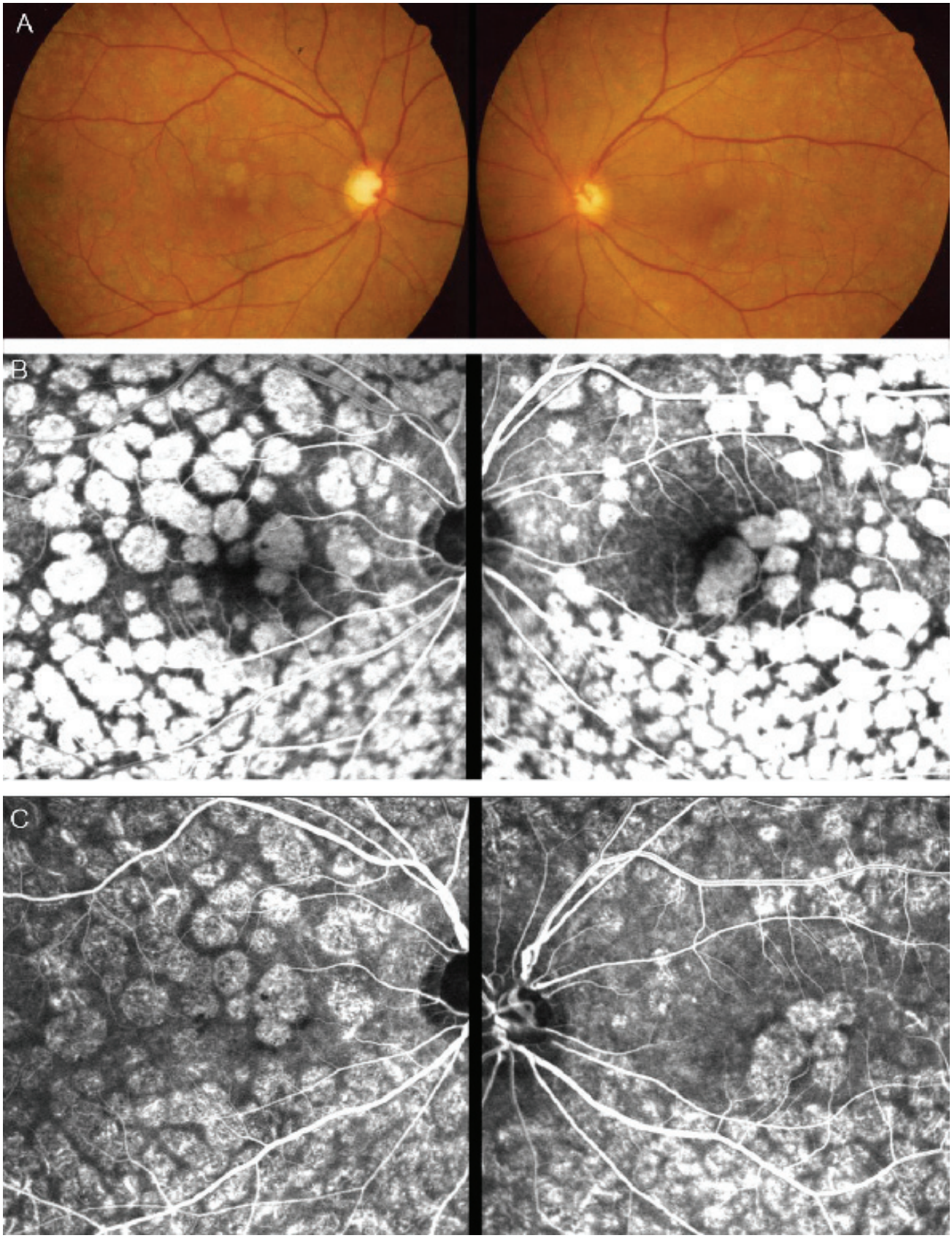

Fig. 2. Two months later. (A) Fundus photograph. (B) Fluorescein angiography. (C) Indocyanin green angiography. Both the number and size of multiple round patches increased.

There are reports of BDUMP without such characteristic features. Wu S et al.[9] reported a case of BDUMP without choroidal lesions. Similar to the case reported by Wu S et al.[9], our case only showed retinal pigment epithelium change. As they mentioned before, such finding is not a result of direct effect of uveal melanocytic proliferation but rather selective loss of retinal pigment cells due to paraneoplastic process. Saito et al.[8] reported a case of BDUMP related to CAR which showed response to steroid therapy. CAR seems to respond to corticosteroid treatment, but BDUMP usually does not. In this case, changes of ERG and visual field corresponded with CAR finding. Therefore, authors considered the possibility of the case being related to CAR and used triamcinolone acetonide 40 mg subtenon injection instead of systemic steroid considering the patient's general condition. However, the patient expired due to deteriorating general condition and hence, we couldn't detect anti-retinal antibody to diagnose CAR and the effectiveness of steroid therapy could not be confirmed.

We report a case of visual paraneoplastic syndrome which showed nummular loss of the pigment epithelial cells which distinguishes the clinical component of bilateral diffuse uveal melanocytic proliferation.

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