Case Report

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Bilateral Congenital Grouped Pigmentation of the Retina in a Patient with Left Superior Rectus Muscle Palsy

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Key Words

Congenital grouped pigmentation

Bear track spots

Abstract

A 39-year-old man with left superior rectus muscle palsy had many sharply circumscribed, variable-sized pigmented spots in both fundi. His visual acuity, visual fields, and color vision appeared normal bilaterally. The electroretinographic responses were also normal. His family members were unaffected by pigmented spots in the retina. We believe that the bilateral congenital grouped pigmentation of the retina found in our patient may be rare.

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Introduction

Congenital grouped pigmentation of the retina, or bear track spots, is a rare congenital anomaly characterized by many sharply circumscribed, pigmented spots of variable size and no visual dysfunction [4]. This anomaly is reportedly unilateral in most patients [2,4] although bilateral cases of familial occurrence have been reported [1, 6]. We recently examined a man with this rare anomaly in both fundi.

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Case Report

A 39-year-old man complained of diplopia in upward gaze. The patient's past medical history was unremarkable: he had never experienced night blindness, serious ocular disease, prolonged diarrhea, rectal bleeding, or tumors of the bowel. His family history was also unremarkable. His visual acuity was 1.5 OU. The intraocular pressures were 14 mm Hg OU. The primary eye position was orthophoric. The right eye movement was undisturbed in all directions, but the left eye movement was interrupted slightly in upward gaze. No ocular pain or proptosis was noted. The eyelids were not ptotic. The corneas, anterior chambers, lenses, and vitreous appeared clear bilaterally. The pupils were isocoric, and pupillary reactions to light were prompt bilaterally. Ophthalmoscopically, many sharply circumscribed, pigmented spots of variable size were visible in the nasal and temporal sectors of

Fig. 1. The right fundus shows many sharply circumscribed, variable-sized pigmented spots in the nasal and temporal sectors.

the right fundus (fig. 1). The retinal lesions had no tail of irregular white depigmentation. The retina appeared ophthalmoscopically normal, except for the pigmented lesions. The optic disc, macula, and retinal vessels of that eye appeared normal. Many small pigmented spots were observed in the temporal sector of the left fundus (fig. 2). The optic disc, macula, and retinal vessels in the left eye appeared normal. Fluorescein angiography disclosed blockage of background fluorescence without leakage. Results of Goldmann visual-field testing, Panel D-15 color vision examination, and electroretinography disclosed no abnormality. Hess test and Bielschowsky head tilting test results disclosed palsy of the left superior rectus muscle.

Fig. 2. The left fundus demonstrates sharply circumscribed, small, pigmented spots in the temporal sector.

Results of laboratory tests including blood cell counts, blood chemistry analyses, *Treponema pallidum* hemagglutination test, serum titers to *Toxoplasma gondii*, serum titers to *Borrelia burgdorferi*, urinalysis, chest X-ray, computed tomography of the brain and orbit, tensilon test, and serum levels of thyroid hormones were negative or within the normal range.

The patient's parents and brother we examined showed no unusual pigmentation of the retina. During the 3-month follow-up, the patient's retinal condition remained unchanged. Three months later, diplopia disappeared spontaneously.

Discussion

itary congenital hypertrophy of the retinal pigment epithelium [7], which is solitary; multiple bilateral pigmented lesions in familial adenomatous polyposis and Gardner syndrome [7], in which multiple intestinal polyps and cancer are found, and the retinal lesions usually have a tail

of irregular white depigmentation; nonspecific reactive proliferation (hyperplasia) of the retinal pigment epithelium [7], which does not show the sectorial arrangement; and bilateral chorioretinitis, in which the retinochoroidal mottling is usually found by ophthalmoscopy, and some of which show the positive results of laboratory tests.

Congenital grouped pigmentation of the retina is reportedly unilateral in most patients [2, 4]. Bilateral conditions, as found in our patient, may be rare, although bilateral cases of familial occurrence have been recently described [1, 6]. The family members of our patient did not have the condition.

Congenital grouped pigmentation of the retina has been reported in association with other pathologic ocular or systemic changes [2, 3]. Our patient also had left superior rectus muscle palsy of unknown cause. The association of the retinal condition and superior rectus muscle palsy in our patient may be unrelated. The retinal condition remained unchanged during the follow-up period, but the muscle palsy disappeared. To our knowledge, the association of these disorders has not been described previously.

We believe that bilateral congenital grouped pigmentation of the retina, as found in our patient, may be rare.

The retinal condition in our described patient was compatible with congenital grouped pigmentation of the retina [2,4]. The retinal lesions in our patient could be differentiated from retinitis pigmentosa [5], in which night blindness, peripheral visual field loss, and reduced to absent rod and cone amplitudes by electroretinography are common; sol-

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