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PECULIAR STREAKS OF THE RETINAL PIGMENT EPITHELIUM SECONDARY TO CHOROIDAL DETACHMENT

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ABSTRACT

A patient that developed peculiar streaks of the retinal pigment epithelium is described. These streaks appear after choroidal detachments of all etiologies, mainly the ones of long duration, and in this case it appeared after an exudative detachment caused by an episode of the Vogt-Koyanagi-Harada Syndrome. These streaks appear as long, black, disconnected lines, curved and angular or forked. Most tend to be meridional in direction and parallel to one another, but some run in a circular fashion. They are more abundant at the equator and many extend anteriorly or posteriorly. They result from hyperplasia of the pigment epithelium wich had accumulated increases formed in the choroid during the detachment.

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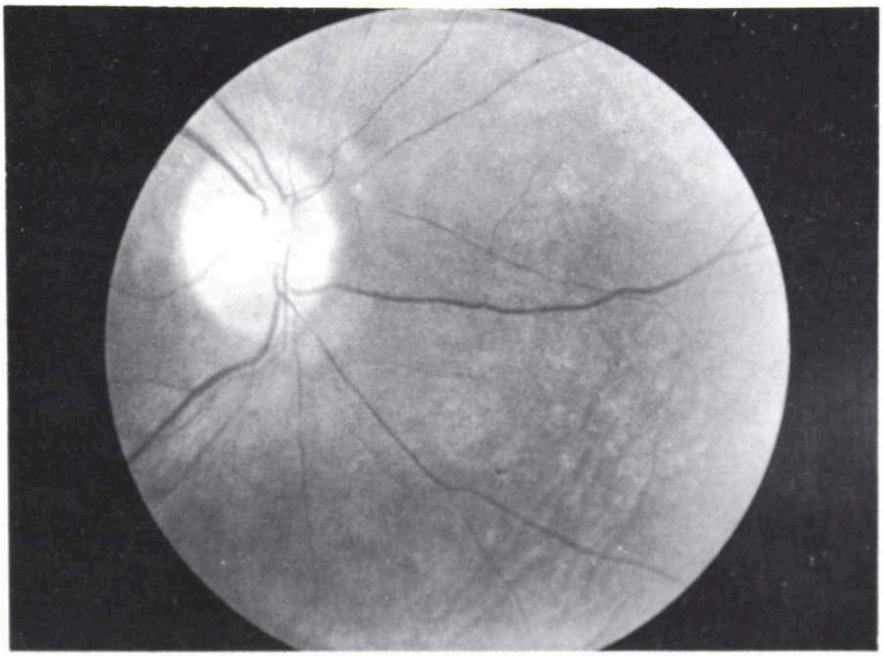
INTRODUCTION

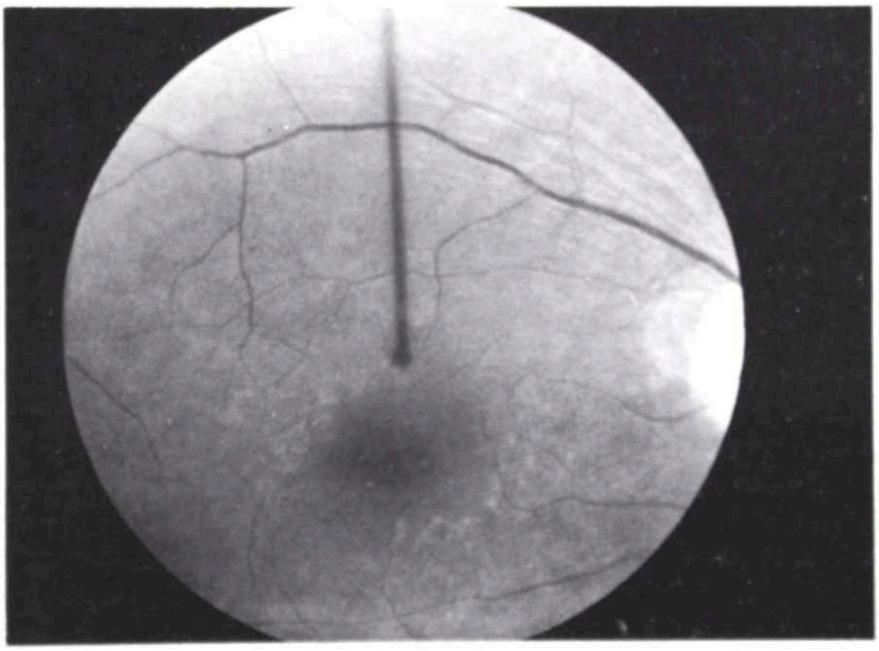
Only a limited number of papers about pigmented streaks secondary to choroidal detachments have appeared in the world's medical literature [10,14]. This finding is probably more common than the small number of publications on this subject might suggest. It is seen after choroidal detachments of all etiologies, mainly those of long duration. This sign has pathognomonic significance since it indicates the presence of a previous detachment of the choroid[5].

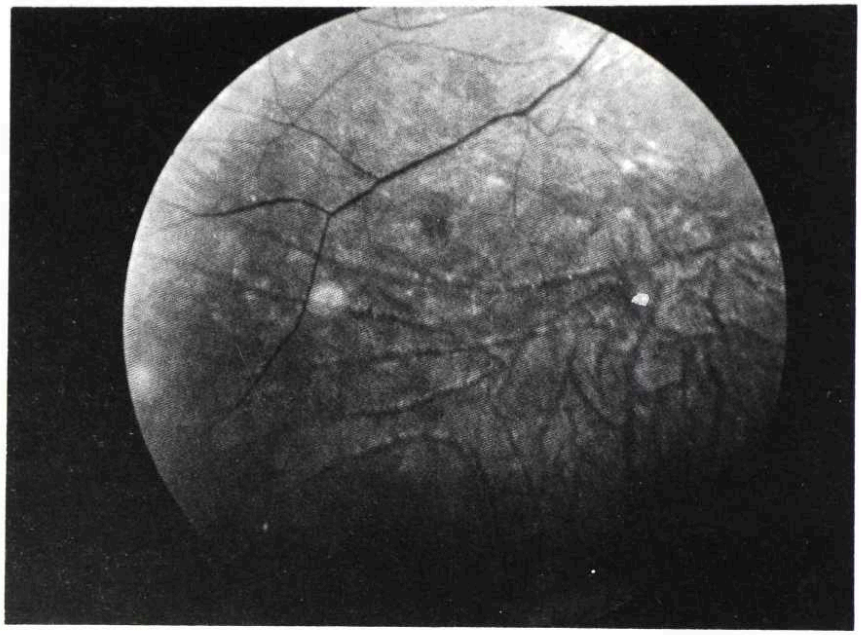
CASE REPORT

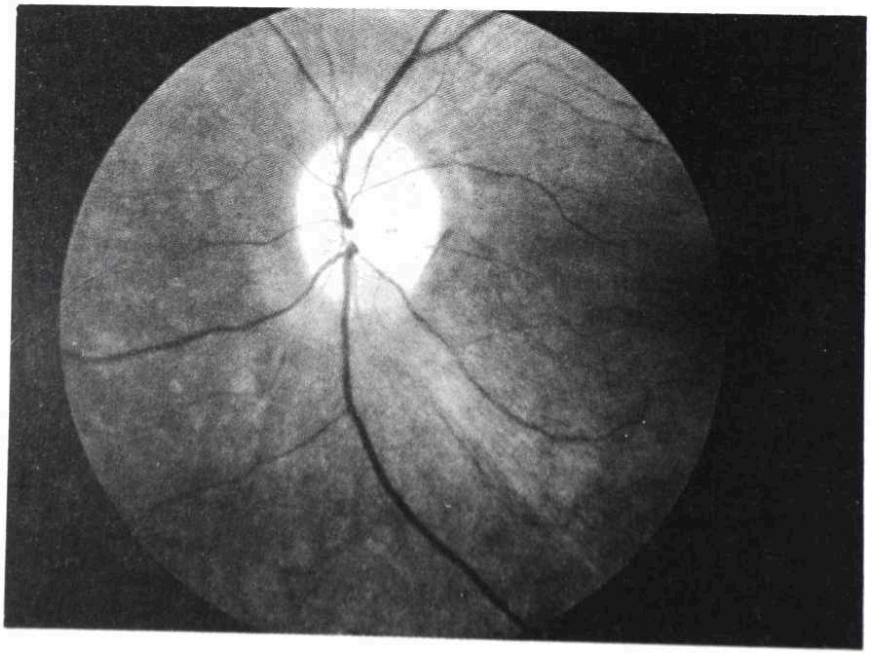
A 47-year-old Iranian female had experienced sudden visual loss in the left eye, followed one month later by envolvement of the righ eye. A diagnosis of Harada's disease wes made while she was living in Iran, and she later a further decrease in vision ocurred. She complained of a hearing problem at that time but reported no history of poliosis or vitiligo, or alopecia.

Nine months after the initial visual loss, she visited the UCLA Clinic with a complaint of sudden painless decrease in vision in the right eye. Her initial visual acuity was light perception OD, 20/400 OS. Examination of the anterior segment showed 1+ flare and cells in the Aqueous of the right eye. There were 1+ vitreous cells and flare in both eyes. Intraocular tension was 18 mmHg in each eye. Funduscopic examination revealed generalized narrowing of the arterioles, pale optic discs, linear whorls of pigmentation posterior to the equator, diffuse areas of deep gliosis, a gray appearance of the retina, and pigment clumpling in the macula (Fig. 1-6). There was an exudative detachment of the right macula. A recurrence of Harada's disease was diagnosed and she was treated with systemic steroids and local atropine. Vision improved to 20/200 00 in the following month. Visual field tests revealed a generalized contraction in both eyes, especially in the temporal fields. Fluorescein angiograms showed pigment epithelial defects around the macula, pigmented streaks disposed in a branching circular and radial fashion around the optic discs, and a striated pattern at the posterior pole (Fig. 7-11). The electro-oculogram examination was performed and there was no light rise in either eye. A photopic electroretinogram was barely recordable in the right eye and was nonrecordable in the left eye. Averaging the photopic responses gave the same result. The scotopic electroretinogram was barely recordable in the right eye and was nonrecordable in the left.









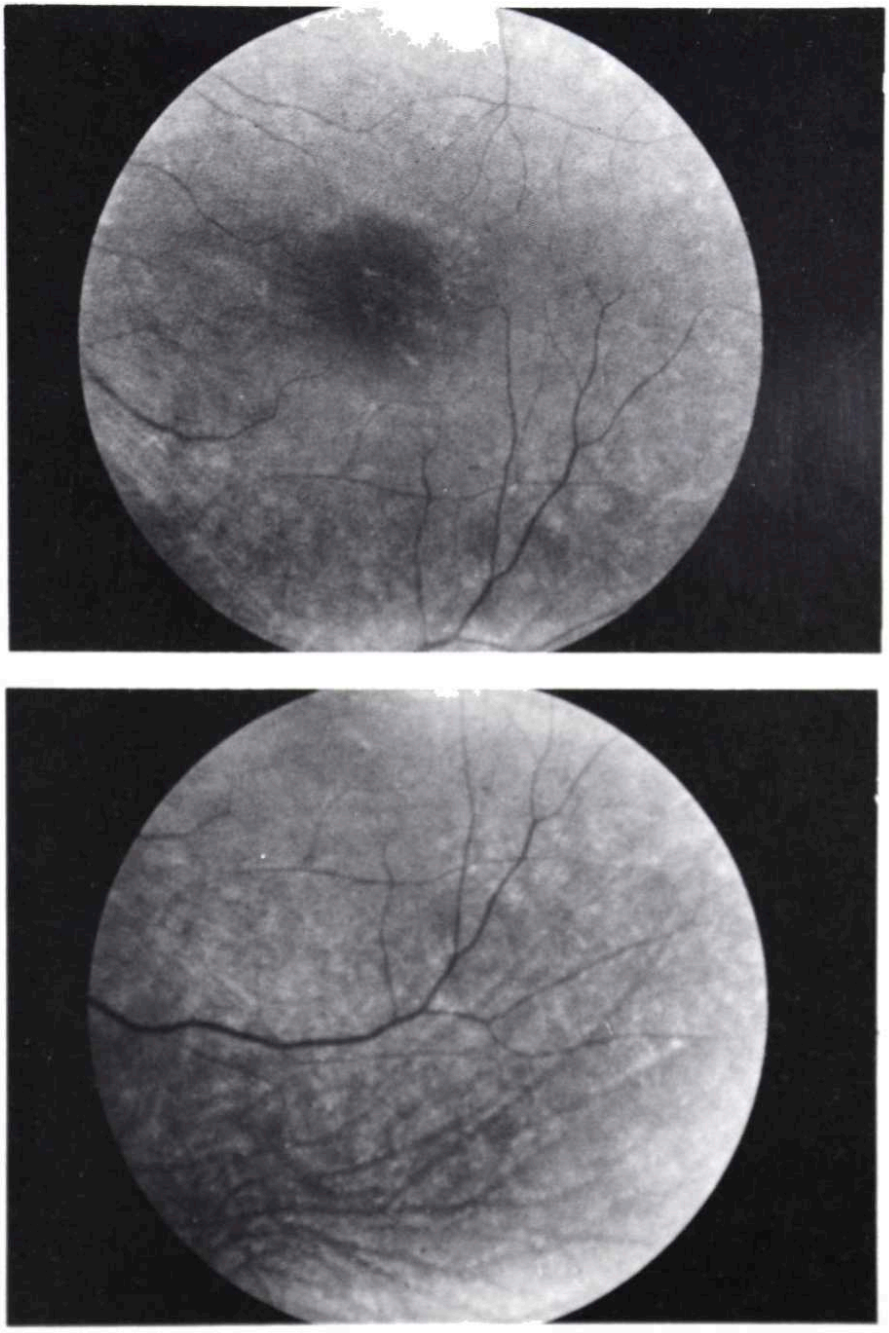
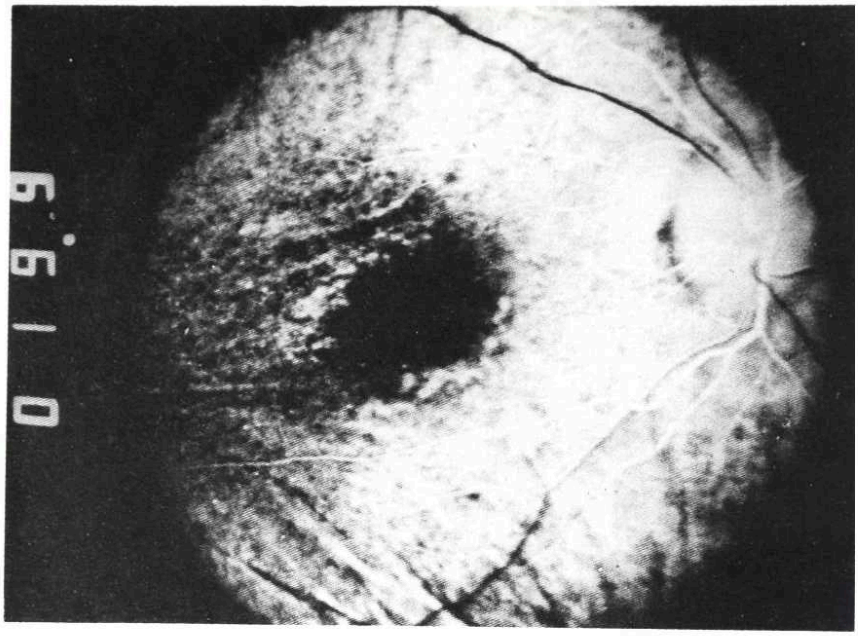


FIGURE 1 THROUGH 6

Fundus aspects show pale optic discs, narrowing of the arterioles, a gray appearance of the retina, pigment clumping at the macula, linear whorls of pigmentation posterior to the equator.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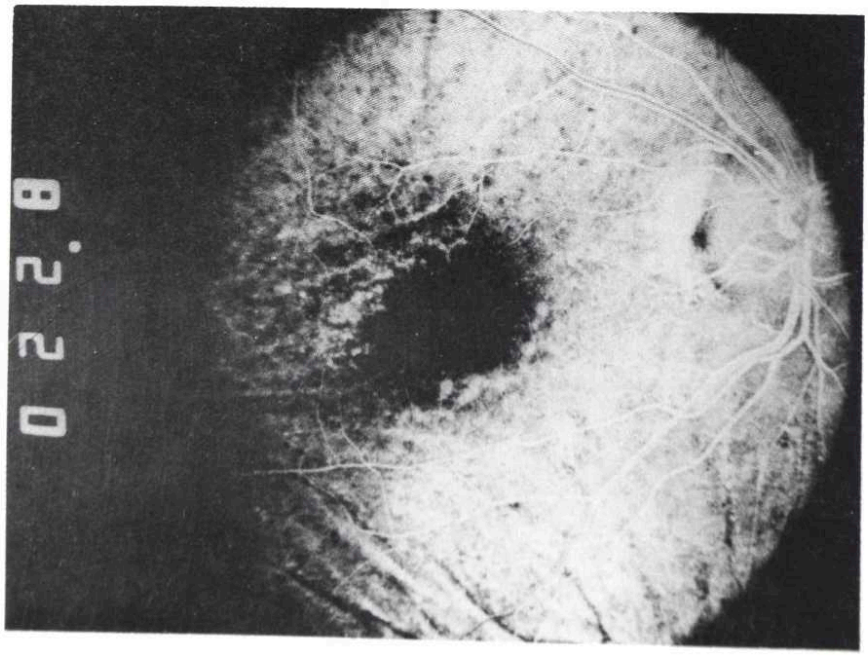
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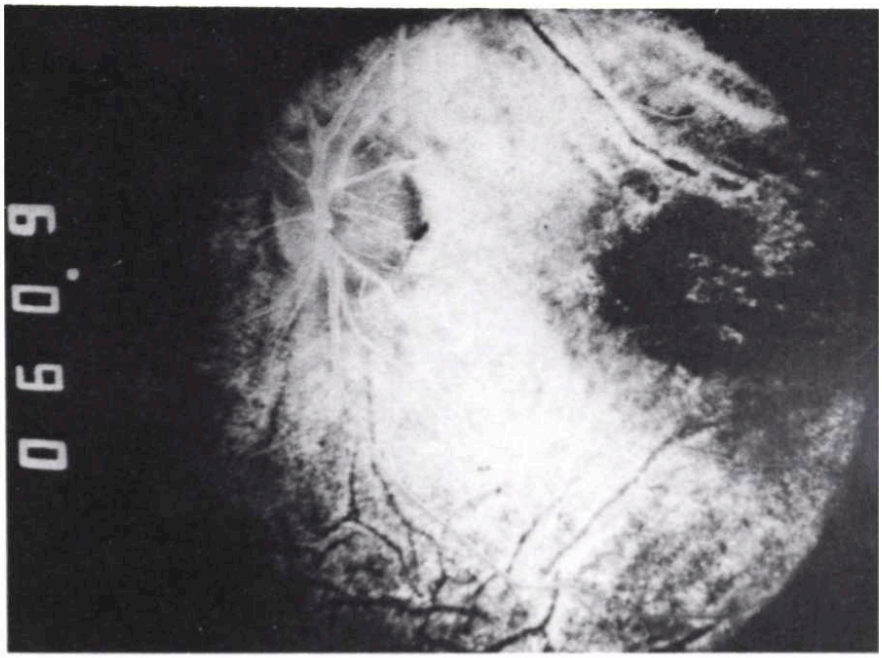
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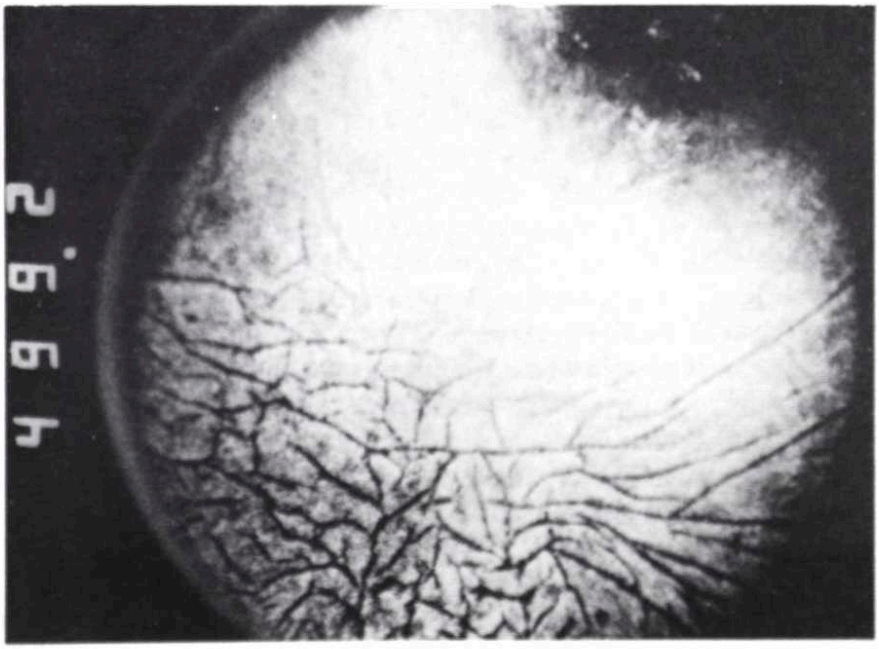
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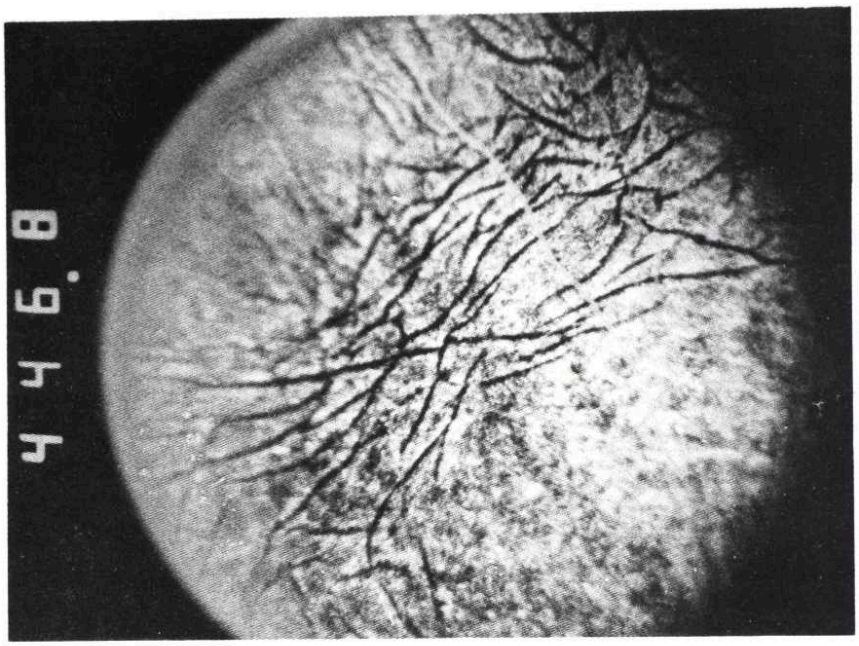

FIGURE 7 THROUGH 11

Fluorescein angiograms show pigment epithelial defects around the macula, pigmented streaks disposed in a branching circular and radial fashion around the optic discs and a striated pattern at the posterior pole.

DISCUSSION

Clinical cases involving pigmented streaks firts were reported by Schur (1913)[10]. Schnaudigel (1913)[9], Plocher (1917)[6], and Fuchs (1918)[3]. Typically these streaks appear as long, black disconnected lines, curved and angular or forked, situated beneath the retinal vessels, granular in composition, and rather thicker than the average retinal vein. They are most abundant at the equator and many extend anteriorly. Seldom do they reach as near as 2 or 3 mm to the optic disc. Most tend to be meridional in direction and parallel to one another, but some run in a circular fashion. In the region of the streaks, the fundus may show pale areas and scattered pigment dots.

Verhoeff (1931)[15] first described their microscopic appearance: ridgelike thickenings of the pigment epithelium with cells containing pigment granules that retain the spindle form seen in normal pigment epithelium. Verhoeff proposed that the streaks resulted from hyperplasia of the pigment epithelium which had accumulated in creases formed in the choroid during detachment.

The generally accepted explanation for this phenomenon is based on the difference in rigidity between the retina and the choroid. When the choroid detaches, it is forced into folds together with the retina. The outer parts of the choroid and retina, being relatively soft, yield to the compression usually without folding further, but the Bruch's membrane with the choriocapillaris and the inner surface of the retina, owing to their greater rigidity, are forced into rounded folds associated with outward creases. If the creases persist long enough, pigment cells accumulate in some of them and appear as pigmented streaks. They have been seen after a detachment that lasted for eight days, but normally a longer period is required for these typical dark lines to become visible[1]. They are easier to see when the detachment has resolved, but they also can be seen while the detachment persists.

The pigmented streaks have been described mainly in postoperative choroidal detachments and after globe injuries. The case reported here is apparently the first in which pigmented streaks were seen after exudative detachment due to Harada's disease. Probably all types of choroidal detachment are potentially able to propitiate the development of these streaks.

Pigmented streaks have been confused with angioid streaks. Spaeth and De Long (1944)[12] described a case of blunt trauma to the eye with a broomstick followed ten days later by choridal detachment. Later, typical pigmented streaks were seen although the authors called them angioid streaks. Differential diagnosis should consider the streaks described by Siegrist in 1899[11]. This kind of streak is rare and is supposed to be due to pressure that arteroesclerotic vessels of the choroid exert on the pigment epithelium, stimulating its proliferation.

The pigment disturbance that commonly occurs in the Vogt-Koyanagi-Harada syndrome is described as a diffuse atrophy of the pigment epithelium followed in late stages of the disease by the appearance of irregular spots of pigment at the posterior pole. The pigment distribution seen in these case is irregular and does not follow the linear pattern seen in the pigment streaks.

Although alopecia, vitiligo, and poliosis were not seen in the present case (being reported), the iridocyclitis and the fundus appearance, which include an exudative retinal detachment, narrowing of blood vessels, pigment disturbances, optic disc pallor, and areas of retinal gliosis, were typical of the Vogt-Koyanagi-Harada syndrome. Also, the dysacusia and mental disturbances which the patient exhibited support that diagnosis. With steroid treatment there was some improvement of vision in the right eye, which is normally seen in this disease. As we did not see this patient at the initial episode when she was in Iran,

we oculd not follow the development of her pigmented streaks and cannot comment about the lengh of time it took for them to form. Plocher[6] affirmed that the presence or pigmented streaks is pathognomonic of a previous choroidal detachment. Although we found no choroidal detachment when we initially examined the patient, it is known that cilio-choroidal effusions are common in patients with Vogt-Koyanagi-Harada syndrome and this patient very probably had this finding at the time of her initial episode in Iran.

Streaks of the retinal pigment epithelium certainly do not occur in every case of cilio-choroidal detachment, and the cases reported in the literature demonstrate that extent and duration of the detachment are determining factors in the formation of pigmented streaks. Meticulous follow-up examinations of patients presenting with choroidal detachments of all causes, could probably dosclose more cases of this interesting finding.

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