Bilateral spontaneously reattached rhegmatogenous retinal detachment. Case report and differential diagnosis with pigmentary retinopathies

ABSTRACT

Background: Dark pigmentation of the ocular fundus presents in degenerative diseases such as retinitis pigmentosa. This disease must be distinguished from other diseases whose evolution is not progressive in order to estimate the functional prognosis of the patient. We undertook this study to analyze the features that distinguish spontaneously reattached retinal detachment from other causes of ocular fundus pigmentation in order to identify it even in bilateral cases.

Clinical case: We present the case of a female with chronic visual loss who was referred for evaluation with the diagnosis of a pigmented retinopathy. Clinical exploration ruled out causes such as retinitis pigmentosa, retinal inflammatory diseases or trauma. Based on the clinical features, topography of pigmentation and information provided by electroretinography, a diagnosis of bilateral spontaneous reattachment of rhegmatogenous retinal detachment was made. Clinical features of this entity are discussed as well as the diagnostic approach to distinguish it from other pigment retinopathies.

Conclusion: Clinical features of spontaneously reattached retinal detachment allow the explorer to distinguish it from other causes of bilateral pigmentation, despite presenting bilaterally. Because the prognosis of the attached retina is better than that of a degenerative disease, the correct diagnosis facilitates rehabilitation.

Key words: pigment retinopathy, retina, retinal detachment.
INTRODUCTION

The retinal pigment epithelium is the outermost layer of the retina, which is attached to the choroid, one of the vascular layers of the eye. When the retinal pigment epithelium develops hyperplasia as a secondary change to some pathological process, the ophthalmoscopic appearance of the fundus of the eye changes from orange to brown and, eventually, to a black color due to the melanin. This dark pigmentation in the fundus of the eye appears in different “pigmentary” retinopathies, the best known being degeneration of the photoreceptors called “retinosis pigmentosa.”

Retinosis pigmentosa is a progressive degenerative process that is necessary to be distinguished from other causes that induce hyperplasia of the retinal pigment epithelium, like some inflammatory diseases (rubella, syphilis), extensive trauma of the retina, medication toxicity or nonprogressive pigmentary retinopathies such as paravenous pigmentary atrophy or congenital hypertrophy of the pigmentary epithelium of the retina. Retinal detachment with spontaneous resolution also courses with accumulation of pigment in the area that is reattached.

When the spontaneously resolved retinal detachment is unilateral, the differential diagnosis is made easily because unilateral retinosis pigmentosa is rare and it is less common to find patients with bilateral pigmentation secondary to inflammation or trauma that usually affects only one eye. Although retinal detachment can occur in both eyes, bilateral spontaneous resolution is rare.

CLINICAL CASE

A 71-year-old female patient with extensive retinal pigmentation in both eyes attended our clinic. She reported visual loss that was long-standing in her right eye and that had started 5 years ago in her left eye; the patient did not remember having had nictalopia. There was a family history of diabetes, but not of vision-imparing ocular diseases. The patient had a cataract extraction without intraocular lens in the right eye at the age of 19, and phacoemulsification with intraocular lens at the age of 16; she had had T2DM during 10 years, which was treated with oral hypoglycemics and arterial hypertension that was treated with beta-blockers. There was no history of trauma, ocular inflammation or treatments with medications associated with retinal toxicity that cause pigmentation.

On the ophthalmic evaluation, visual acuity was no light perception in the right eye and 20/150 that did not improve with glasses in the left eye; the refraction was +15.00 = -2.00 × 30° in the right eye and +2.25 = -2.00 × 135° in the left. There was an afferent pupillary defect in the right eye, intraocular pressure was 18 mmHg on the right side and 19 on the left; the corneas were transparent, there was not inflammation in the anterior chamber, the iris was normal and there were no synechiae. The right eye was aphakic and the left had a stable posterior chamber intraocular lens; the anterior vitreous was syneretic and did not have any pigment.

The right ocular fundus had an orange-yellow disc with well-defined margins, a 0.3 disc diameter cup, central vascular emergence and a 2:3 arteriovenous ratio. There was atrophy in the peri-papillary pigment epithelium and in the macula. The retina was attached, with bone spicules between meridians 11 and 1, and pigment clusters between meridians 1 and 11. There were three lines of subretinal gliosis: one superior to the disc, another in the 2 meridian and one in the 3 meridian (Figure 1).

In the left eye fundus, the yellow-orange disc had well-defined margins, with a 0.3 disc di-
ameter cup, central vascular emergence and a 2:3 arteriovenous ratio. There was atrophy of the pigment epithelium in the macula and the retina was attached. From meridians 1 to 10 there were pigment clusters in the retina and three lines of subretinal gliosis: one below the inferior border of the disc, another below the inferior temporal arcade and one more on the inferior periphery. The superior retina between meridians 10 to 1 was not pigmented and had microaneurysms and dot and blot hemorrhages (Figure 2).

The patient had abnormal retinal pigment in the entire fundus of the right eye, but the superior left eye fundus had no pigment changes. Although the bilaterality suggested a pigmentary retinopathy, the absence of pigment in the superior left retina was not consistent with degeneration affecting all photoreceptors. Although sector forms of retinitis pigmentosa exist, it is more common to find the changes in a smaller area than that of the healthy retina than the inverse.

The optic disc was not pale and, although the extent of the pigmentation made difficult evaluating the vascular attenuation in the equatorial retina, the emergent main vessels of the disc and those in the superior retina of the left eye did not show thinning. The absence of signs in the anterior segment ruled out inflammatory disorders; there was no history of ocular trauma to relate the pigmentary change.

The multiple lines of subretinal gliosis was not compatible with the diagnosis of retinitis pigmentosa. Although some inflammatory diseases may cause subretinal fibrosis, gliosis is in retinal detachments with spontaneous resolution. The subretinal appears in zones where the subretinal fluid stops, in a manner similar to the demarcation lines in patients with idiopathic juvenile retinal dialysis.

The electroretinogram reported absence of binocular photopic response and scotopic response only in the left eye. The visual-evoked potentials showed response only in the left eye with prolonged P100 wave latency.

**Figure 1.** Fundus of the right eye. Dark color pigmentation between meridians 1 and 11 and bone spicules between meridians 1 and 11. The optic disc was not pale and there were three lines of subretinal gliosis.

**Figure 2.** Fundus of the left eye. Dark color pigmentation in meridians 1 to 10. The optic disc was not pale, and there were spot microaneurysms and hemorrhages in the nonpigmented superior retina.
Because of the absence of other signs of retinitis pigmentosa and inflammatory signs, the existence of multiple lines of subretinal gliosis, and the electroretinogram characteristics, the final diagnosis was bilateral spontaneously resolved rhegmatogenous retinal detachment. The patient also had mild non-proliferative diabetic retinopathy in the retina without pigmentation.

**DISCUSSION**

Retinal detachment is a disease that can cause blindness where the retina separates from the retinal pigment epithelium to which it is normally attached. In the rhegmatogenous type there is a retinal break that allows communication between the vitreous cavity and the subretinal space that is virtual under physiological conditions. In addition to retinal break, rhegmatogenous retinal detachment is associated with vitreal traction and movement of the vitreous gel fluid.\(^7\)

Morbidity is significant when the macula is affected before the detachment is diagnosed. In symptomatic rhegmatogenous retinal detachments, surgical treatment is indicated.\(^8\) The principles of surgery for treatment of rhegmatogenous retinal detachments include creating a chorioretinal adhesion to block all retinal holes, removing retinal traction and draining the subretinal fluid in some cases.\(^9\)

In 1981, Cantrill\(^10\) described cases of rhegmatogenous retinal detachments with spontaneous resolution. Although this evolution is more frequently reported in patients with macular holes,\(^11-13\) Cho et al.\(^14\) in 2007 published a case series whose causative lesion was not found at the macula. In that study, retinal dystrophies and uveitis were excluded as the cause of the pigmentation due to the unilaterality and location of the lesions.

The patient in the case presented had bilateral involvement and did not have signs of inflammation in the anterior segment; therefore, the diagnosis of uveitis was ruled out.

In the 15 eyes reported by Cho et al.\(^14\) there were two specific characteristics: diffuse accumulation of pigment, retinal pigment epithelium atrophy, or both and a convex margin of the lesion. Only in one eye did the pigment accumulation encompass the entire fundus of the eye, the average extension was 6 meridians; 11/15 eyes had subretinal gliosis (73.3%).

In the patient whose case is reported here, pigment clusters extended throughout the fundus of the right eye and in 7 meridians on the left. Due to the history of prior surgery at 19 years of age and the extent of the disorders, it is probable that the retinal detachment would have been complete and of longer time in the right eye, which could explain the lack of light perception.

The median visual acuity in the study by Cho et al.\(^14\) was 20/50 (range of hand motion 20/20); in the five eyes in which the visual was worse than 5/200 there was pigment that involved the macula. In the functional eye of the patient in this case, visual acuity was 20/150, but there was no pigment in the macula.

In the case here presented there were no causative lesions identified in either of the eyes, but in the study by Cho et al. only in one of the eyes was a causative lesion found.\(^14\)

Lorenzo et al. proposed that detachment of the posterior vitreous could free the tractional component in retinal detachment and contribute to the resolution of the detachment. Also, the detached vitreous could occlude the causative lesion and limit the detachment.\(^15\) In a report of three cases with spontaneous resolution of the retinal detachment in which a causative lesion was identified, Chung et al.\(^16\) detected that one had partial detachment of the vitreous. The area
where the detachment with spontaneous resolution was located had the vitreous attached.

In non-Caucasian patients, retinal detachments associated with inferotemporal dialysis also could have a spontaneous resolution, principally because of the inferior location of the disease. The spontaneously reattached retina could accumulate pigmentation due to hyperplasia of the retinal pigment epithelium. This pigmentation that outlines the extent of the detachment is known as the line of demarcation and is reported in 60% of the cases.17

Although it is common for the retinal dialysis to be bilateral, it is uncommon that they cause total retinal detachments, or the subretinal gliosis found in rhegmatogenous retinal detachments with spontaneous resolution.18 The patient presented in this case had multiple lines of subretinal gliosis that would have corresponded to different stages of progression of the rhegmatogenous detachment.

Spontaneous resolution of a rhegmatogenous detachment of the retina is uncommon, but even more rare is for this evolution to be bilateral. In the study by Cho et al.,14 four of five patients had retinal detachment of the contralateral eye, but in none was a spontaneous resolution reported.

An additional possibility would be that the patient would have had retinitis pigmentosa to which was added a rhegmatogenous retinal detachment. The cases reported with this condition have shown other characteristics of retinitis pigmentosa such as a pale optic disc and vascular attenuation19 or paravascular distribution of the pigment in the shape of bone spicules,20 but not the subretinal gliosis found in the rhegmatogenous retinal detachments of spontaneous resolution.

Patients with retinitis pigmentosa that affects the periphery usually have poor night vision due to the condition of the rods. One individual with degeneration of the photoreceptors (to the extent that the patient of the case reported had) would have perceived visual deficiency under low light conditions. Night blindness orients towards the diagnosis of retinitis pigmentosa; although absence of this symptom could rule out retinitis pigmentosa, the patient in the case presented had cataracts from an early age, and the night vision impairment might not have been evident if the degeneration of the photoreceptors had begun at a later date.

In retinitis pigmentosa the electroretinogram typically shows signal loss of rods (scotopic) and of cones (photopic), although the disorder of the rods tends to predominate.21 The patient presented in this case had scotopic response in the left eye and multiple lines of demarcation of the retina, but not the remaining clinical characteristics of retinitis pigmentosa, for which reason this diagnostic possibility was ruled out.

Although the visual prognosis of the patient was poor in the right eye, the functional retina of the left eye did not have the expectation that it would have had if she had been diagnosed with retinitis pigmentosa, a disease in which retinal degeneration is progressive. The diagnosis of retinal detachment with spontaneous bilateral resolution implied that there was function in the unaffected retina of the left eye, and that visual loss related to an already existing diabetic retinopathy should be prevented.

In conclusion, the clinical characteristics of rhegmatogenous retinal detachment with spontaneous resolution allow this condition to be distinguished from other diseases that have retinal pigmentation. The prognosis of a retina that has not detached is better than for a degenerative disease, because it allows for visual rehabilitation that can be facilitated by an accurate diagnosis.
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REFERENCES