Congenital hypertrophy of retinal pigment epithelium

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DESCRIPTION

A 45-year-old man was referred to the ocular oncology service for evaluation due to a black asymptomatic lesion in his right eye. His medical history included hyperlipidemia and high myopia. On clinical examination, a flat black lesion that looked like a flower was observed (figure 1A,B). The lesion did not resemble melanoma. Moreover, ultrasound examination confirmed that the lesion was flat. The lesion showed hypoautofluorescence at the dark parts that correlate with the known histopathologic evidence of lack of lipofuscin in the retinal pigment epithelium of congenital hypertrophy of retinal pigment epithelium (CHRPE) and mild hyperautofluorescence that correlate with

Figure 1 Flower of congenital hypertrophy of retinal pigment epithelium (CHRPE). (A) Colour fundus photography—a black flat lesion in the peripheral retina resembling a flower. (B) An autofluorescence image—the lesion shows hypoautofluorescence that correlates with the known histopathologic evidence of lack of lipofuscin in the retinal pigment epithelium of CHRPE. The lacunae show mild hyperautofluorescence that correlates with scleral autofluorescence. (C) Optical coherence tomography (OCT) with a raster line on the lesion area—a loss of the external retinal layers is depicted. (D) OCT with a raster line on the lacunae area—no retina is seen in the lacunae area.

Patient's perspective

Feel well, arrive to annually follow-up.

Learning points

- ▶ Classic congenital hypertrophy of retinal pigment epithelium (CHRPE) is usually unifocal, benign, pigmented lesion located at the level of the retinal pigment epithelium.
- ▶ On optical coherence tomography, CHRPE is flat with irregular external retinal layers in the pigmented portion and absent external retinal layers in the lacunae area.
- ▶ All patients with CHRPE should be asked about colon cancer and diseases in their family.

scleral autofluorescence at the lacunae (figure 1B). In optical coherence tomography, a loss of the external retinal layers was seen in the area of the lesion (figure 1C) and no retina was seen in the lacunae' area (figure 1D). The diagnosis was confirmed as CHRPE.

In revised anamnesis, no positive familial history of polyps, Gardner syndrome or colon cancer was noted.

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