Int Ophthalmol (2019) 39:441–444
https://doi.org/10.1007/s10792-017-0806-x

PHOTO ESSAY

Choroidal osteoma and pattern dystrophy of retinal pigment epithelium

Vinod Kumar

Received: 21 July 2017 / Accepted: 11 December 2017 / Published online: 19 December 2017

Abstract
Purpose To describe co-occurrence of choroidal osteoma in a patient with pattern dystrophy of retinal pigment epithelium.
Methods Clinical case report
Results A young female presented with decreased vision in the right eye due to decalcification of choroidal osteoma. Multimodal imaging including fundus autofluorescence, fluorescein angiography and optical coherence tomography showed features of pattern dystrophy of retinal pigment epithelium that simulates the Stargardt disease.
Conclusions This co-occurrence of choroidal osteoma and pattern dystrophy is likely to be incidental. Multimodal imaging may help in differentiating pattern dystrophy of retinal pigment epithelium that simulates Stargardt disease from Stargardt disease.

Keywords Choroidal osteoma · Pattern dystrophy · Retinal pigment epithelium · Multimodal imaging

A 30-year-old female presented with gradual progressive decrease of vision in her right eye for 6 months. Systemic and family history was non-contributory. Best-corrected visual acuity was 20/80 in the right eye and 20/20 in the left eye with − 1 dioptre sphere in both the eyes. Anterior segment examination was normal in both eyes. Dilated fundus examination of the right eye showed a whitish orange choroidal mass on the supero-temporal aspect of the optic disc (Fig. 1a), which extended superiorly and nasally (Fig. 2a). In addition, multiple greyish-yellow globular flecks were seen at the posterior pole of both the eyes sparing the centre of the macula (Fig. 1a, b). Short wave fundus autofluorescence (SWAF) of the right eye showed diffuse hypoautofluorescence corresponding to the choroidal lesion (Fig. 1c). The retinal flecks were hypo- as well as hyperautofluorescent (Fig. 1c, d). Ultrasound B scans of the right eye showed highly reflective lesion adjacent to the optic nerve shadow (Fig. 2b). A high spike was seen on corresponding A scan, and the lesion caused shadowing underneath simulating a characteristic double optic nerve head appearance. Fundus fluorescein angiography (FFA) showed diffuse hyperfluorescence corresponding to the choroidal lesion (Fig. 3a). All the flecks were seen to be hyperfluorescent on FFA, and 'dark choroid' was not appreciated. Swept source optical coherence tomography (SS-OCT) horizontal scans through the fovea of both eyes were obtained. SS-OCT of right eye (Fig. 4a) showed hyper-reflective choroidal lesion with disruption of overlying outer retinal layers that extended up to fovea. SS-OCT of left eye (Fig. 4b) showed flecks as hyper-reflective

V. Kumar (✉)
Dr Rajendra Prasad Centre for Ophthalmic Sciences, All India Institute of Medical Sciences, New Delhi 110091, India
e-mail: drvinod_agg@yahoo.com

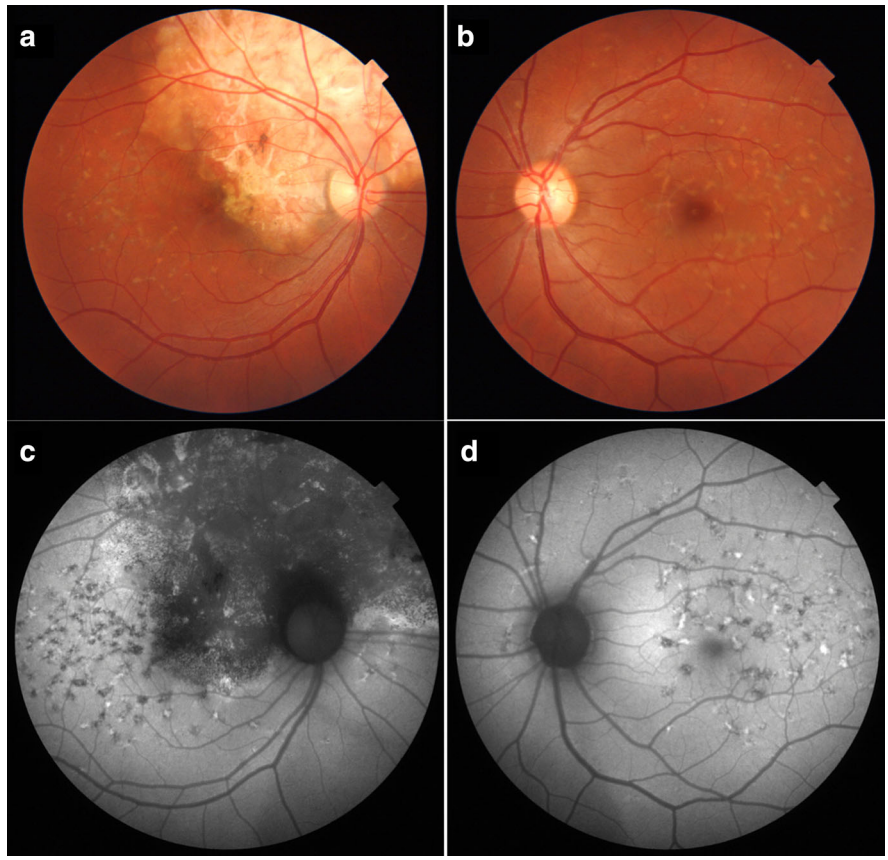

Fig. 1 Colour fundus photographs of right (**a**) and left (**b**) eyes showing choroidal osteoma in the right eye and globular flecks in both the eyes. Autofluorescence imaging shows osteoma as hypoautofluorescent, and flecks are hypo- as well as hyperautofluorescent (**c**, **d**)

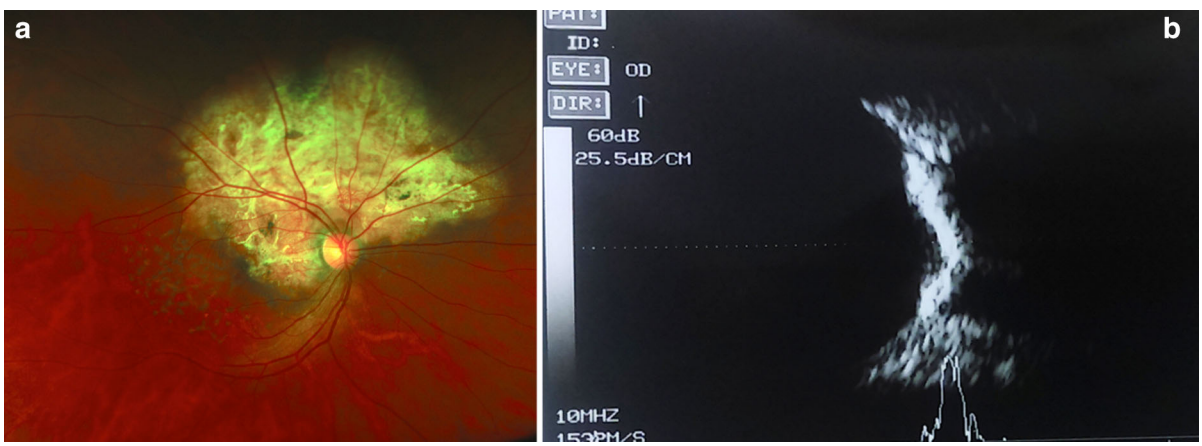

Fig. 2 Wide field photograph shows extent of osteoma (**a**). Ultrasound B scan shows double optic nerve appearance because of osteoma (**b**)

material on the upper surface of retinal pigment epithelium (RPE). These flecks breached the ellipsoid zone and extended up to the external limiting membrane (arrow, 4b). Based on the clinical picture, a diagnosis of pattern dystrophy simulating Stargardt disease along with choroidal osteoma in the right eye was made. The patient was apprised of the situation and was advised regular follow-up and Amsler grid charting at home.

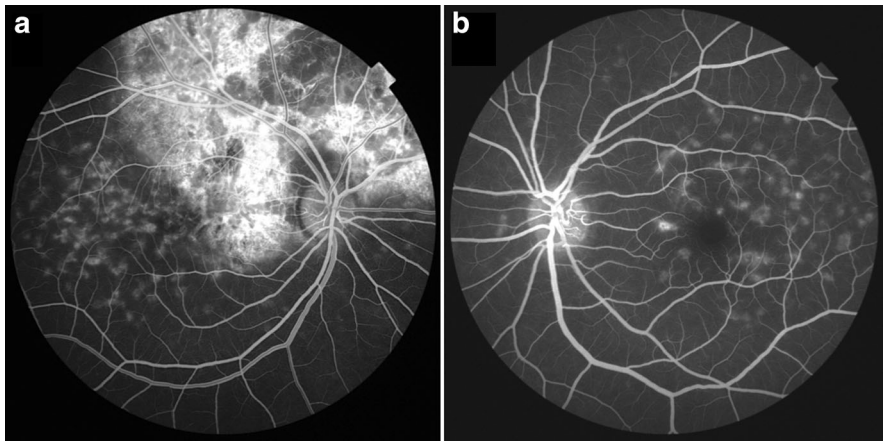

Fig. 3 Fundus fluorescein angiography shows hyperfluorescent globular flecks (**a**, **b**) and hyperfluorescence corresponding to the osteoma (**a**)

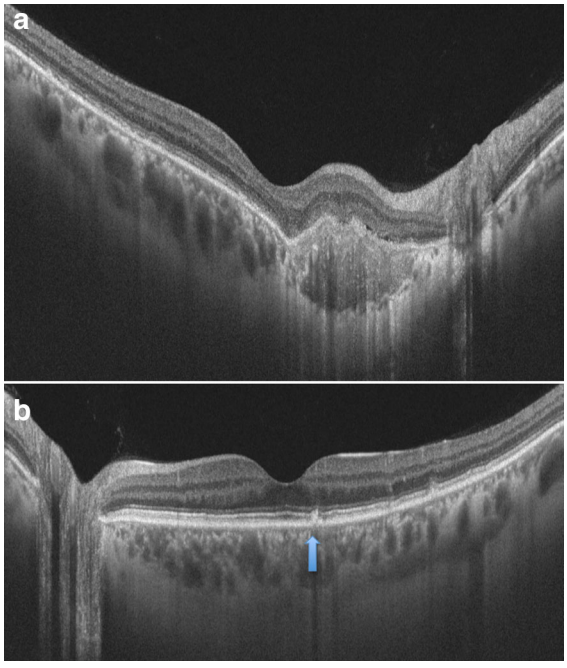

Fig. 4 SS-OCT scan of right eye shows hyper-reflective choroidal osteoma in the right eye (**a**) and flecks located above the RPE in the left eye (arrow, **b**)

Comment This case highlights coexistence of two rare diseases in a single patient. Such an association has not been described before and is likely to be coincidental. Choroidal osteoma tends to decalcify over a period of time. Its decalcification is associated with loss of overlying RPE and outer retinal layers [1]. This results in the gradual visual loss if the macula is involved. The poor visual acuity in the right eye could be attributed to loss of RPE in the macular area. SWAF is an excellent tool for assessment in such situations, and hypoautofluorescence over the area of choroidal osteoma indicates decalcification and loss of RPE.

The flecks seen in this patient were more globular as compared to pisciform flecks of Stargardt disease. Their characteristic appearance as hyper-reflective material on the surface of RPE has been described before [2]. The combination of globular flecks, absence of dark choroid and sparing of central macula suggested the diagnosis of pattern dystrophy.

Interestingly both pattern dystrophy and choroidal osteoma are associated with increased incidence of choroidal neovascularisation (CNV) [3, 4]. A close follow-up is therefore recommended in such a patient for early detection of CNV, which responds well to intra-vitreal injections of anti-vascular endothelial-derived growth factor in both the conditions [5, 6].

To conclude, this report highlights coexistence of two rare anomalies and their potential implications.

Compliance with ethical standards

Conflict of interest The author declares that he has no conflict of interest.

Ethical approval All procedures performed in this report were in accordance with the institutional guidelines and with the 1964 Helsinki Declaration and its later amendments.

Informed consent Informed consent was obtained.

References

1. Aylward GW, Chang TS, Pautler SE, Gass JDM (1998) A long-term follow-up of choroidal osteoma. Arch Ophthalmol 116:1337–1341

2. Kumar V, Kumawat D (2017) Multimodal imaging in a case of butterfly pattern dystrophy of retinal pigment epithelium. Int Ophthalmol. http://doi.org/10.1007/s10792-017-0497-3
3. Shields CL, Sun H, Demirci H et al (2005) Factors predictive of tumor growth, tumor decalcification, choroidal neovascularization, and visual outcome in 74 eyes with choroidal osteoma. Arch Ophthalmol 123:1658–1666
4. Marano F, Deutman AF, Leys A, Aandekerk AL (2000) Hereditary retinal dystrophies and choroidal neovascularization. Graefes Arch Clin Exp Ophthalmol 238:760–764
5. Parodi MB, Iacono P, Cascavilla M, Zucchiatti I, Kontadakis DS, Bandello F (2010) Intravitreal bevacizumab for subfoveal choroidal neovascularization associated with pattern dystrophy. Invest Ophthalmol Vis Sci 51:4358–4361
6. Narayanan R, Shah VA (2008) Intravitreal bevacizumab in the management of choroidal neovascular membrane secondary to choroidal osteoma. Eur J Ophthalmol 18:466