Korean J Ophthalmol 2015;29(4):282-283
http://dx.doi.org/10.3341/kjo.2015.29.4.282

# A Case of Congenital Simple Hamartoma of the Retinal Pigment Epithelium and Coats' Disease in the Same Eye

Dear Editor,

Congenital simple hamartoma of the retinal pigment epithelium (CSHRPE) is a rare retinal pigment epithelium (RPE) tumor, incidentally diagnosed in asymptomatic children and young adults. It usually appears as a small, dark pigmented, nodular mass within the macula [1]. Coats' disease is an idiopathic, progressive retinopathy with retinal vascular telangiectasia, exudation, and frequent retinal detachment occurring more commonly in male children [2]. Herein, we describe a male diagnosed with CSHRPE and Coats' disease simultaneously in the same eye, which has not been previously reported.

A 23-year-old Korean male was referred to our hospital with a history of reduced vision in his right eye for 4 years. He had no medical or ophthalmic disease history. On examination, the best-corrected visual acuity was 20 / 63 in the right eye and 20 / 20 in the left eye. The left eye was unremarkable. Fundus examination of the right eye revealed a small, well-circumscribed, dark pigmented, minimally elevated nodular mass involving the fovea, as well as retinal vascular telangiectasia, multiple aneurysms, and tortuous dilated vessels with yellowish-white exudation in the temporal midperipheral retina (Fig. 1A). Fluorescein angiography (FA) revealed persistent blockage of fluorescence at the fovea due to the mass lesion, and subtle leakage from its surroundings in the late phase. Early hyperfluorescence due to retinal vascular telangiectasia and hypofluorescence due to exudation and capillary drop-out were present in the temporal midperipheral retina (Fig. 1B and 1C). Spectral domain optical coherence tomography (OCT) showed well-demarcated retinal elevation with intense hyper-reflectivity on its inner surface, and complete optical shadowing of the outer retina at the fovea (Fig. 1D).

With the diagnosis of CSHRPE and Coats' disease, focal argon laser photocoagulation was performed around the abnormal vascular lesion temporal to the macula. The best-corrected visual acuity of the right eye was 20 / 40 and both lesions were stable at the 24-month follow-up.

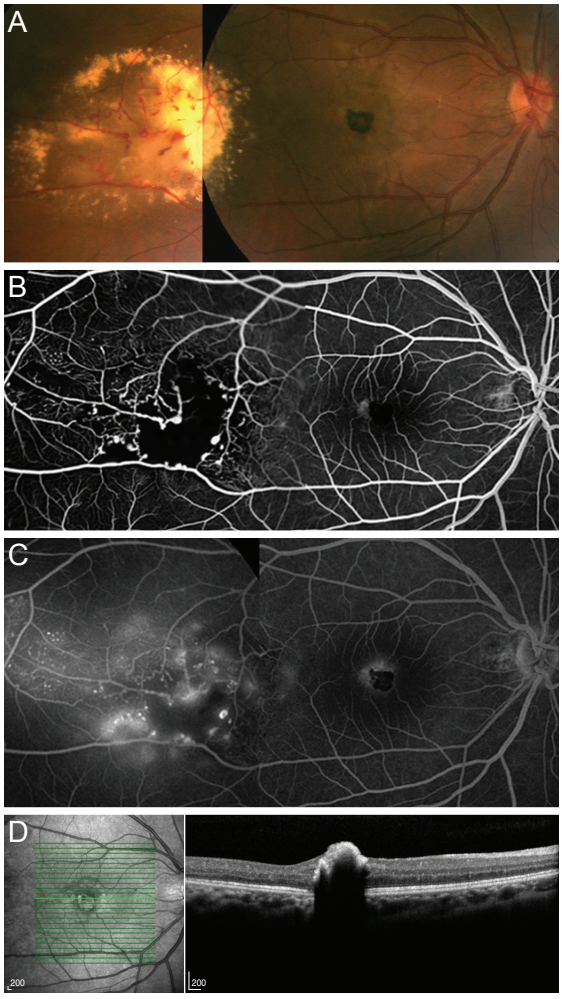


**Fig. 1.** (A) Fundus photograph at initial examination of the right eye shows a small, well-circumscribed, darkly pigmented mass involving the fovea. Telangiectasia of retinal vessels and yellowish-white exudation are also seen in the temporal midperipheral retina. (B) Early phase of fluorescein angiography shows blockage of fluorescence in the fovea due to a mass lesion and a halo of hyperfluorescence around the foveal mass lesion. In the temporal midperipheral retina, aneurysmal dilatation of retinal vessels and hypofluorescence due to exudation and capillary drop-out are seen. (C) Late phase of fluorescein angiography shows subtle leakage around the foveal mass lesion and vascular leakage in the temporal midperipheral retina. (D) Spectral domain optical coherence tomography of the right eye shows a highly reflective, well-defined retinal elevation with complete optical shadowing of the fovea.

CSHRPE is a rare RPE tumor which appears as a dark pigmented, nodular mass with well-circumscribed margins near the fovea. In previous cases, FA showed early nonfluorescence and a late halo of fluorescence at the border of the mass. OCT showed bright reflectivity of the anterior

surface, deep optical shadowing, and definite vertical margins with full thickness retinal involvement [1,3]. These FA and OCT findings were similar to our case.

Khurana et al. [4] reported a case of Coats' disease with subfoveal nodule in an untreated eye. They explained that in Coats' disease, there is preferential accumulation of exudation in the macular area, often remote from the main areas of telangiectasia. Subsequently, subretinal exudation may be transformed into dense fibrous tissue, and can take the shape of a nodule with fibrous metaplasia of RPE. Jumper et al. [5] regarded this subfoveal nodule as macular fibrosis. In their report of 47 cases of Coats' disease, macular fibrosis was present at presentation or developed during the course of follow-up in 11 patients (23%). The macular fibrosis formed in an area of previous dense exudate accumulation and included the fovea in all cases. Temporal peripheral retinal telangiectasia was found in all cases with macular fibrosis. Intraretinal vascular anastamosis within the area of fibrosis was found in 7 of the 11 eyes and hyperpigmentation was present at the point of anastomosis in six eyes. FA showed minimal hypofluorescence before formation of fibrosis and hyperfluorescence and late leakage after formation of macular fibrosis due to intraretinal vascular anastomosis. In our case, the foveal lesion differed from subfoveal nodule or macular fibrosis in morphology, color, and FA findings. Therefore, the diagnosis of foveal lesion in our case was CSHRPE rather than foveal nodule or macular fibrosis in Coats' disease.

The relationship between CSHRPE and Coats' disease is unknown, and a combined case of the two different ocular diseases in the same eye has not been previously reported. Because they show different clinical features, separate management of the two ocular conditions was required. In the present case, focal laser photocoagulation was applied only to the lesion of Coats' disease, and the two different lesions were observed in stable states at the 24-month follow-up. Further careful continuous observation would be helpful for a better understanding of CSHRPE and Coats' disease, especially due to the rare incidences of these diseases.

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## Conflict of Interest

No potential conflict of interest relevant to this article was reported.

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