Indocyanine Green Angiography of Retinal Astrocytomas Associated with Tuberous Sclerosis

Nilufer Koak, Ali Osman Saatci, Suleyman Kaynak, Mehmet Hilmi Ergin, Guray Craymngil

Department of Ophthalmology, Dokuz Eylul University, Izmir-Turkey

As small astrocytic hamartomas can sometimes be missed during routine ophthalmoscopy in patients with tuberous sclerosis, fluorescein and/or indocyanine green angiography maybe more helpful in identifying such small astrocytomas. In fluorescein angiography, astrocytomas show gradually increasing hyperfluorescence due to the vascular permeability of astrocytomas. In indocyanine green angiography, astrocytomas appear hypocyanescent, most prominently during the late phases. We report the indocyanine green angiographic findings of retinal astrocytomas in a patient with tuberous sclerosis.

Key words: astrocytic hamartoma, fluorescein angiography, indocyanine green angiography, retina, tuberous sclerosis

INTRODUCTION

Tuberous sclerosis is a rare hereditary phakomatosis first reported by Bourneville in 1880. Vogt described its classic clinical triad of epilepsy, mental retardation and adenoma sebaceum in 1908. The characteristic ocular finding in tuberous sclerosis is astrocytic hamartoma of the retina or optic disc. Other retinal findings include retinal pigmentary disturbances ranging from hyperpigmented areas to punched out hypopigmented areas at the posterior pole or midperiphery.

We report the indocyanine green angiographic findings of retinal astrocytomas in a patient with tuberous sclerosis.

CASE REPORT

A 14-year-old girl was referred to us with the established diagnosis of tuberous sclerosis from age 2 years. Her systemic examination revealed hypomelanotic macules, bilateral renal angiomyolipoma, and shagreen patch. Mitral valve prolapse was noted in echocardiography. MRI studies revealed multiple nodules (≈ 0.5 × 0.5 cm) in the subependymal region. CT showed bilateral periventricular subependymal calcific nodules. She had ten healthy siblings.

On ophthalmological examination, her best uncorrected visual acuity was 20/20 OU. Ocular motility was full. Slit-lamp examination was unremarkable. Intraocular pressures were within normal OU range. On dilated fundus examination there were three retinal astrocytomas in OD (Fig.1) located at inferior to disc, nasal to disc and superotemporal macula. There were two retinal astrocytomas located at inferior retina and inferior macula in OS.
(Fig. 2). All astrocytomas were flat, translucent, soft-appearing lesions of approximately 1/2 disc-diameter in size.

Fluorescein and indocyanine green angiographies were performed by Heidelberg scanning laser ophthalmoscope. While fluorescein angiography demonstrated gradually increasing hyperfluorescence throughout the angiography (Fig. 3A, B), indocyanine green angiography displayed hypocyanescence due to masking of the astrocytomas which became more distinct during the late phases (Fig. 4A, B).

** Fig. 1. ** Right eye, color photo showing two of the three astrocytomas.

** Fig. 2. ** Left eye, color photo showing one of the two astrocytomas.

** Fig. 3. ** A: Right eye, venous phase of fluorescein angiogram showing hyperfluorescence of two of the three astrocytomas. B: Left eye, venous phase of fluorescein angiogram, composite picture obtained by Heidelberg scanning laser ophthalmoscope showing hyperfluorescence of both astrocytomas.

**DISCUSSION**

Retinal or optic nerve astrocytomas have been well-described in tuberous sclerosis. In a large
astrocytic hamartoma is essentially a benign condition, complications may occur infrequently such as vitreous hemorrhage, retinal vascular abnormalities (including telangiectasia, neovascularization and exudation) and vitreous seeding.º

To the best of our knowledge, indocyanine green angiographic features of retinal astrocytoma have not been reported before. In our case, retinal astrocytomas showed hypocyanesence throughout the indocyanine green angiography which is most likely - due to the masking effect of astrocytomas. However, as tumors may show varying degrees of vascularization and as the tumor’s blood vessels are permeable to fluorescein there is increasing hyperfluorescence during fluorescein angiography.º

Though indocyanine green angiography does not yield additional information and retinal astrocytomas can be well - detected in fundus fluorescein angiography, it may be better to perform indocyanine green angiography in addition to fluorescein angiography to avoid missing any astrocytomas in routine ophthalmologic examination.

REFERENCES