Imaging Findings in Tuberous Sclerosis With Multiple Retinal Astrocytic Hamartomas

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Figure 1. Fundus photograph (top) of the right eye shows retinal hamartomas superior and inferior to the optic disc in a patient with tuberous sclerosis. These hamartomas are glial tumors of the retinal nerve fiber layer that arise from retinal astrocytes and appear as yellowish-gray, well-circumscribed, elevated nodular lesions. Retinal astrocytic hamartomas are most frequently associated with tuberous sclerosis, but may also be found rarely in patients with neurofibromatosis. Solitary, large, retinal astrocytomas can also occur in the absence of any systemic manifestations as an isolated presentation. Fluorescein angiography (bottom) shows staining of the lesions with hyperfluorescence in early phase and no leakage of dye in late phases of angiogram.

Figure 2. Fundus photograph (top) of the left eye shows a large retinal astrocytic hamartoma in the macula superior to the fovea and a second one above the optic disc. The macular hamartoma has a "mulberry-like" appearance. Fluorescein angiography (bottom) shows staining of the lesions.
Figure 3. Spectral optical coherence tomography (OCT) (Cirrus HD-OCT; Carl Zeiss Meditec, Dublin, CA) of retinal astrocytic hamartomas in the right eye reveals elevation of the retina with thickening of the retinal nerve fiber layer by the tumor. The OCT image demonstrates a gradual transition from normal retina into an optically moderate to hyperreflective mass, with retinal disorganization and subtle shadowing of the tissues under the tumor. Despite its reflectivity, only subtle shadowing was observed, which could possibly be because of the translucency of the tumor.

Figure 4. Spectral optical coherence tomography (OCT) (Cirrus HD-OCT; Carl Zeiss Meditec, Dublin, CA) of retinal hamartoma superior to the optic disc in the left eye shows a retinal tumor localized within the nerve fiber layer with the outer retinal layers intact. However, the macular hamartoma does not show thickening or elevation of the retina. Retinal astrocytic hamartomas are commonly elevated, but can also appear flat. Gradual transition from normal retina into an optically moderate to hyperreflective mass with retinal disorganization and subtle shadowing can also be observed in the OCT images.

REFERENCES

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