

# An Incidental Retinal Hemangioblastoma in a case of Central Serous Chorioretinopathy

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**Abstract:** Retinal hemangioblastoma (capillary hemangioma) is a benign vascular tumor of the retina or optic nerve head. It can be sporadic or associated with von Hippel-Lindau (VHL) disease. Three distinct forms of juxta papillary capillary hemangioblastomas have been described, including exophytic, endophytic, and sessile forms. An endophytic tumor appears as an orange-red lesion. However, exophytic and sessile forms are difficult to diagnose as they do not have the characteristic appearance of hemangioblastoma. They are misdiagnosed as papillitis, unilateral papilledema, choroidal hemangioma, choroiditis, or choroidal neovascularization. We report a case of incidentally detected juxta papillary hemangioblastoma on fundus fluorescein angiography (FFA) and optical coherence tomography angiography (OCTA) in a case of central serous chorioretinopathy (CSCR).

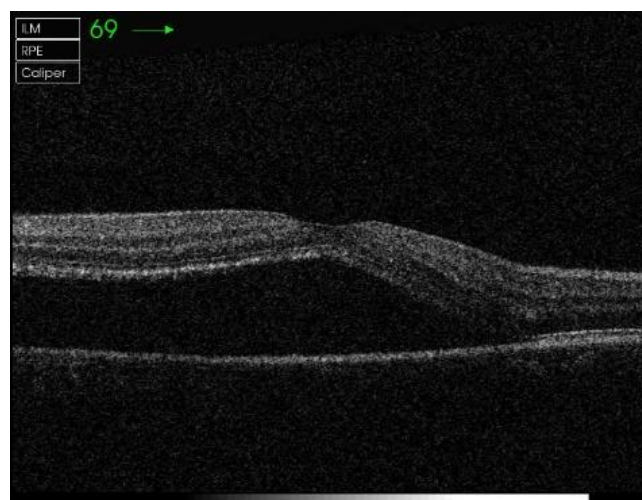
**Keywords:** Retinal hemangioblastoma, FFA, OCTA, CSCR

## 1. Introduction

Retinal hemangioblastoma (capillary hemangioma) is a benign vascular tumor of the retina or optic nerve head. It can be sporadic, or a component of von Hippel-Lindau (VHL) disease<sup>[1]</sup>. VHL syndrome is an autosomal dominant disorder including retinal capillary hemangioblastoma, central nervous system hemangioblastoma, pheochromocytoma, hypernephroma, and several other tumors<sup>[2]</sup>. The locus for the VHL gene is on chromosome 3 (3p25-26), and inactivation of a tumor suppressor gene appears to play a key role<sup>[2]</sup>. The mean age at diagnosis is 18 years for patients with VHL and 36 years for those without VHL. Juxta papillary hemangioblastomas arise on the optic nerve head or within the juxta papillary region. Because of their location they can be misdiagnosed as papilledema, or papillitis and when subtle, are easily missed<sup>[1]</sup>. Fundus fluorescein angiography (FFA) and optical coherence angiography (OCTA) help diagnose such clinically subtle tumors.

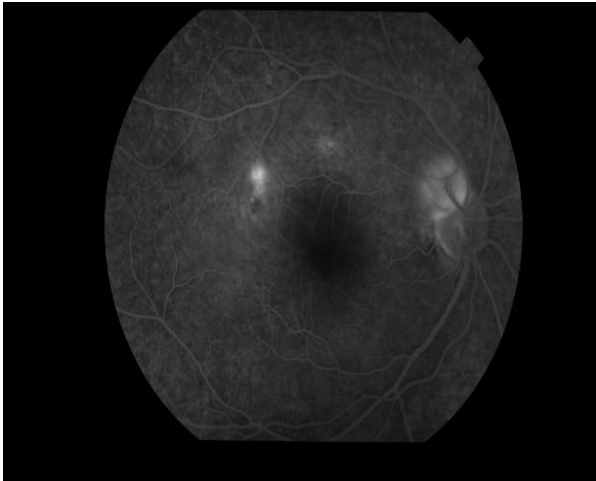
## 2. Case Report

A 36-year-old male presented to our center with the blurring of vision in his right eye for ten days. Ocular examination showed best-corrected visual acuity 20/30 in his right eye and 20/20 in his left eye. Slit-lamp bio microscopy with a 90-diopter lens showed a well-delineated serous detachment at the macula. Optical coherence tomography showed focal pigment epithelium detachment (PED) with neurosensory detachment (NSD) at macula suggestive of central serous chorioretinopathy (CSCR) (Fig. 1).



**Figure 1:** Spectral-domain optical coherence tomography through macula showing neurosensory detachment.

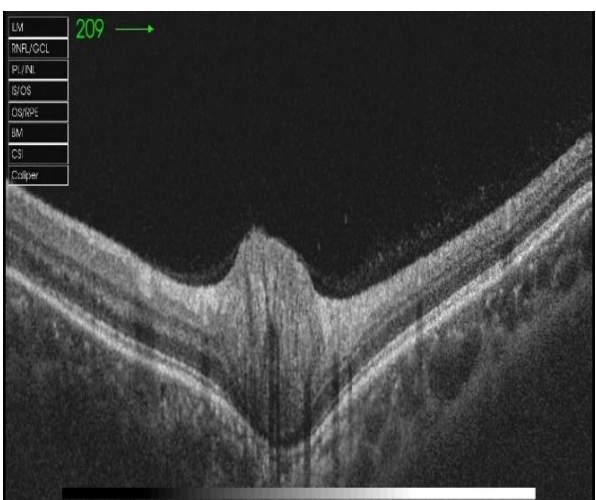
Topical dorzolamide drops QID was given and he was advised to follow-up after a month. At follow-up visit OCT was repeated. The height of the neurosensory detachment was reduced but not completely resolved. After 15 days, FFA was performed. A small hyperfluorescent focus was noted superotemporal to the fovea in early frames which increased in size and intensity in late frames suggestive of leakage (Fig 2). In the arterial phase, hyperfluorescence was noted superotemporal to the disc which increased in intensity in late frames (Fig 2). On slit-lamp bio microscopy, a greyish lesion was noted superotemporal to the disc which was missed earlier (Fig. 3). Optical coherence tomography angiography (OCTA) showed hyperreflective inner layers with thickening corresponding to the FFA lesion (Fig 4). Superficial slab showed a branching network of vessels (Fig 5). Based on multimodal imaging a diagnosis of juxta papillary hemangioblastoma was made.



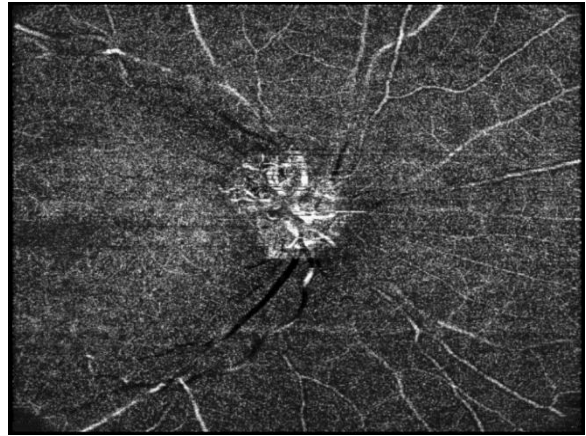
**Figure 2:** Fundus fluorescein angiography of the right eye showing leakage superotemporal to the fovea and at the disc



**Figure 3:** Fundus photo showing greyish lesion superotemporal to disc.



**Figure 4:** Spectral domain optical coherence tomography angiography through disc showing hyperreflective inner layers with thickening



**Figure 5:** Superficial slab showing a branching network of vessels

### 3. Discussion

Early lesions of retinal hemangioblastoma can be missed on slit-lamp biomicroscopy and indirect ophthalmoscopy<sup>[3]</sup>. FFA can identify subclinical lesions thereby enabling early evaluation and management. In our case, the lesion was identified incidentally on FFA which was missed on clinical examination with slit-lamp biomicroscopy. Optical coherence tomography angiography (OCTA) confirmed our diagnosis. Endophytic juxta papillary retinal hemangioblastoma is an orange-red and can be easily diagnosed. However, exophytic or sessile forms are difficult to diagnose and can be misdiagnosed as papillitis, unilateral papilledema, choroidal neovascular membrane, or choroidal hemangioma<sup>[4]</sup>. Fundus fluorescein angiography can identify the vascular nature of the tumor and aids in the diagnosis. The exact location and depth of the tumor and vascularity are well delineated on OCTA. Hence, OCTA can be used as a non-invasive tool in the diagnosis of retinal hemangioblastoma.

Retinal hemangioblastomas are associated with macular edema, exudation, tractional or exudative retinal detachment. Management depends on tumor size, location, and associated complications<sup>[5]</sup>. Tumors associated with VHL syndrome require aggressive treatment. Several treatments have been proposed such as laser photocoagulation, photodynamic therapy (PDT), brachytherapy, transpupillary thermotherapy, intravitreal anti-VEGF, and surgical excision. Small asymptomatic tumors can be observed<sup>[6],[7]</sup>. Juxta papillary tumors are difficult to treat because of their location<sup>[7]</sup>. Anti-VEGF agents alone or in combination with PDT are a good option. Multimodal imaging would be helpful in diagnosis of subtle juxta papillary retinal capillary hemangioblastoma.

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