Brief Report

Submacular sclerosing capillary hemangioblastoma

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A R T I C L E   I N F O

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A B S T R A C T

Purpose: To report a case of submacular cystic lesion that turned out to be a sclerosing capillary hemangio-
blastoma. The retinal capillary hemangioblastoma may arise as a part of von Hippel-Lindau syndrome 1 however,
they may occur as an isolated entity 2, but submacular capillary hemangioblastoma has never been reported.
Observations: A case of a 56-year-old female who presented with a right yellow submacular lesion. The mass was
excised via pars-plana vitrectomy and histopathological assessment revealed a submacular sclerosing capillary
hemangioblastoma.
Conclusion: and importance: The capillary hemangioblastoma is a benign vascular tumor that may arise sub-
macularly and is not necessarily associated with von Hippel-Lindau disease. It is difficult to be differentiated
clinically from other submacular lesions but the possibility should be considered.

1. Case report

A fifty-six-year old female presented with a recent diminution of
vision in the right eye of 3-months duration. The clinical evaluation
revealed normal anterior segment, intact ocular motility and the visual
acuity was counting fingers 27.5 inches. The fundus examination re-
vealed a large round elevated submacular lesion with well-defined
borders (Fig. 1). The Optical coherence tomography (OCT) revealed a
well-defined large subfoveal lesion elevating the overlying retina
(Fig. 2). The fluorescein angiography showed a hyperfluorescent lesion
with a mild late leakage (Fig. 3). The examination of the left eye was
unremarkable.

An excisional biopsy was planned via a 23 gauge pars plana vi-
trectomy. Intraoperatively, a yellow round lesion was identified. It was
well-defined with no firm adhesions to the optic nerve or the sur-
rounding structures. It was possible to deliver the mass as one unit by
enlarging the sclerotomy incision. It was a cystic soft yellowish-brown
compressible lesion measuring 1 × 0.5 centimeters. The postoperative
course was uneventful and the visual acuity of the patient improved to
0.1.

Histopathological examination demonstrated an angiomatous lesion
formed of small vascular spaces lined by flat endothelial cells together
with vacuolated stromal cells, however, there was no evidence of
atypia, necrosis, specific infection or malignancy in all the examined
sections (Fig. 4). A diagnosis of submacular sclerosing capillary he-
angioblastoma was confirmed. The patient was referred to the med-
ical oncology department for further systematic evaluation to exclude
von-Hippel Lindau syndrome after pathology indicated the lesion to be
a retinal capillary hemangioblastoma. Neuroimaging, urine analysis,
renal ultrasonography were done and all results were normal.

2. Discussion

The sclerosing capillary angioma was first reported as a rare benign
tumor arising from the lung, originating from pulmonary epithelium,
hence the name 'Pneumocytoma' but it was a hemangioma rather than
a hemangioblastoma.3 The retinal capillary hemangioblastoma is a rare
tumor that may occur as an isolated lesion, but more commonly asso-
ciated with von Hippel-Lindau syndrome.1,2 The tumor can be located
anywhere in the fundus, with juxtapapillary site is common with early
involvement of the macula with leakage and exudation.4 The sub-
macular location of sclerosing capillary hemangioblastoma has not
been reported and to the best of our knowledge, this is the first case
report at this abnormal location. We belive that the lesion originated
from the underlying choroidal vasculature with subsequent sclerosis.

3. Conclusions

The retinal capillary hemangioblastomas are well-differentiated
benign lesions that may affect vision through leakage, exudation and/or
hemorrhage.5 Their presence in the submacular space is unique and has
not been reported before.

The absence of these complications in our case may be related to
sclerosis, however, it still affects vision due to its location with the
disruption of the photoreceptors.

It represents a challenging situation to the surgeon excising this submacular lesion.

The clinical and systemic evaluation must be done to rule out associated von Hippel-Lindau disease.

**Fig. 1.** Fundus photography showing yellow submacular lesion. (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)

**Fig. 2.** Optical Coherence Tomography (OCT) showing submacular lesion elevating the overlying reina.

**Fig. 3.** Fluorescein angiography demonstrated hyperfluorescent lesion with a mild late leakage.

**Fig. 4.** Histopathologic examination showing small vascular spaces lined with flat endothelial cells.

**Intellectual property**

We confirm that we have given due consideration to the protection of intellectual property associated with this work and that there are no impediments to publication, including the timing of publication, with respect to intellectual property. In so doing we confirm that we have followed the regulations of our institutions concerning intellectual property.

**Research ethics**

We further confirm that any aspect of the work covered in this manuscript that has involved human patients has been conducted with the ethical approval of all relevant bodies and that such approvals are acknowledged within the manuscript.

Written consent to publish potentially identifying information, such as details or the case and photographs, was obtained from the patient(s) or their legal guardian(s).

**Authorship**

All listed authors meet the ICMJE criteria. We attest that all authors contributed significantly to the creation of this manuscript, each having fulfilled criteria as established by the ICMJE.
Patient consent

Written consent for publication of personal identifying information including medical record details and photographs was obtained from the patient.

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Conflicts of interest

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Appendix A. Supplementary data

Supplementary data related to this article can be found at http://dx.doi.org/10.1016/j.ajoc.2018.05.010.

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