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Original Research Paper

OPHTHALMOLOGY

A CASE OF UNILATERAL SPORADIC SOLITARY CAPILLARY HEMANGIOBLASTOMA ASSOCIATED WITH CIRCINATE RETINOPATHY – A RARE PRESENTATION.

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ABSTRACT Retinal capillary hemangiomas are benign vascular tumors that arise from the retina or optic disc and those associated with von hippel landau disease occur earlier, more frequently and bilaterally compared to sporadic RCH's. We report below a rare case of solitary sporadic capillary hemangioma in the right eye complicated by circinate retinopathy with an absolutely normal retina of the other eye. A detailed history with thorough clinical and ocular examination with the necessary investigations was done and treatment was initiated. Two months following the treatment, the patient showed an improvement in visual acquity.

KEYWORDS : : Sporadic, capillary hemangioma, von hippel lindau, circinate.

INTRODUCTION: Capillary hemangioblastomas are vascular hamartomas with specific clinical and angiographic features.[1] They may be peripheral or peripapillary in location[1]. They are progressive, have feeder vessels and lead to varying degrees of retinal exudation (circinate retinopathy to exudative retinal detachment).[1] The causes of vision loss from RCH's include exudation from the tumor causing retinal edema, glial proliferation on tumor surface eventually leading to tractional and or exudative retinal detachment.[4]

The diagnosis is primararily clinical; ophthalmoscopically they appear as an orange mass with dilated feeder vessels.[2][3] Retinal capillary hemangiomas are commonly bilateral; according to a large cross sectional study carried out by National Eye Institute, RCH was unilateral in 42% and bilateral in 58%.[5]Von Hippel Lindau syndrome is characterized by RCH (most frequent finding), infratentorial tumors, renal cell carcinomas and pheochromocytomas and is more common with bilateral RCH's.[1]
Histopathology – There are three types of cells of RCH's which include endothelial cells, pericytes and "foamy" stromal cells. Of these, the endothelial cells are fenestrated which provide the basis for exudation which is a characteristic feature of RCH.[6][7](fig : 1)

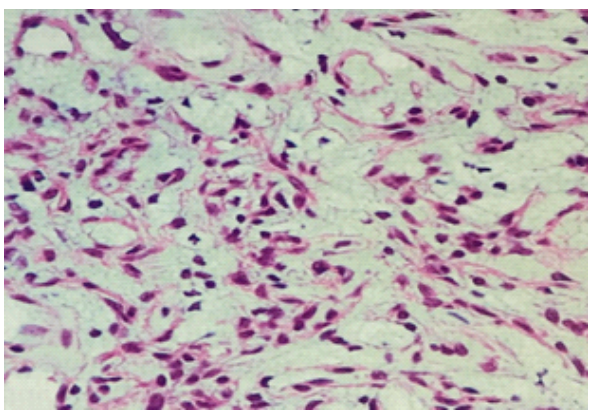


Fig 1: The above image shows many vacuolated foamy stromal cells.

The available treatment options are laser photocoagulation, cryotherapy, radiation, photodynamic therapy or by surgical excision. Argon laser photocoagulation can fully treat retinal capillary hemangiomas of any size in most locations.[8]-[11]
CASE: A 42 year old male came to our OPD complaining of diminution of vision in right eye which was gradual, painless and progressive in nature.

On ocular examination:
1. Both eyes anterior segment: no abnormality detected.

2. Intraocular tension: Right eye – 20.6; left eye -14.6 (measured with applanation tonometer)

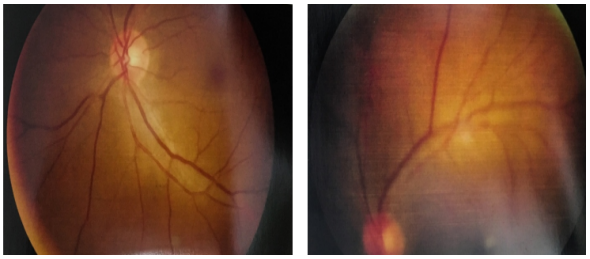
3. Visual acquity: Right eye – Finger counting close to face; Left eye – 6/12 with pinhole improving upto 6/6.

4.Dilated Ophthalmoscopy using Indirect Ophthalmoscope :
Right eye: Disc – size, shape – normal , C:D ratio- 0.3, margins – well-defined

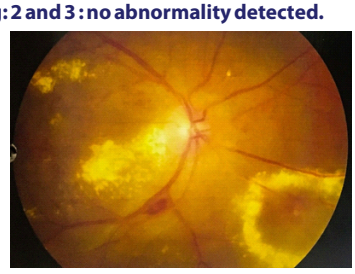
Macula – FR dull
Peripheral retina – Single orange - red colored mass inferior to disc with dilated vessels encroaching towards it.

Multiple hard exudates in the periphery of the retina forming circinate pattern in macular area Left eye : Fundus: no abnormality detected.

5. Fundus photography : (fig : 2, 3 and 4)



Left eye - Fig: 2 and 3 : no abnormality detected.



Right eye - Fig: 4 : Single orange - red colored mass seen inferior to disc with dilated feeder vessels encroaching towards it. Multiple hard exudates in the periphery of the retina forming circinate pattern in macular area.

The following investigations were done in view of Von hippel lindau disease:

CBC, Serum urea and creatinine, Urine routine and microscopy and urine collection for VMAs.

MRI brain and renal USG.
All the above reports were normal.
The patient was treated with a single setting of argon laser photocoagulation.

DISCUSSION: The average age at initial detection of RCH is usually between 15 to 35 years with the mean age for diagnosis of VHL disease being 25 years. This case of RCH is a relatively rarer entity as its unilateral and not associated with VHL. A single RCH has caused severe visual loss producing large extent of exudation involving the macular area. One setting of Argon laser photocoagulation was done which caused a significant improvement in visual acquity over a period of two months from finger counting close to face to 6/60. Regular follow ups of this patient will be required to continue with further line of management and for screening for VHL disease as per the Cambridge screening protocol[12]

CONCLUSION: Prior to the invent of comprehensive screening techniques, the median age of survival of patients with VHL disease was less than 50 years. The causes of death were complications associated with renal cell carcinomas and CNS hemangiomas. Hence, early diagnosis even in eyes with a solitary hemangioma to rule out VHL disease and prompt management with available modern imaging modalities has become the need of hour in order to avoid fatality due to the disease.

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