

Retinal Vasculitis Associated with Vasoproliferative Tumour and Hyperhomocysteinemia: A Case Series

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Abstract

Case1: reported a 31 year old male with complaints of floaters since 15 days with nil systemic history. Both eye BCVA was 6/6 and anterior segment was normal. RE fundus was normal. Patient had hyperemic disc, splinter haemorrhage, well circumscribed elevated lesion with tortuous blood vessels over it with surrounding sub retinal exudation and inferior vitreous haemorrhage was noted. **Case 2:** reported a 22 years male with complaints of gradual painless diminution of vision in right eye for past 7 years. Patient had Hypertension for past 2 months on treatment. Patient had Right eye exotropia of 15 degree with BCVA 6/36 and LE 6/12. He had circumcorneal congestion in both eyes. RE occasional cells and LE cells 2+ with flare 1+. LE fundus showed vitritis 1+, dilated tortuous vessels over the posterior pole, temporal periphery showed pre retinal fibrosis and thick membranes, dilated tortuous feeder vessels with Vaso proliferative tumour was noted in the inferotemporal quadrant and subretinal exudation along vessels were noted. **Case 3:** reported a case 40 year-old female was diagnosed with LE intermediate uveitis + peripheral mass ITQ near the pars plana with vitritis 1+ with vision LE 6/9 N6 and RE was normal with visual acuity of 6/6 N6. LE showed a yellowish mass raising from a snow bank surrounded by vitreous inflammatory snow balls with tortuous fine retinal vessels within the lesion. In all 3 cases fundus fluorescein angiography showed capillary ferning in all quadrants in late venous phase with hyperfluorescent lesion corresponding to the vascular leakage from the tumour. Serum homocysteine level was found elevated. All other uvea work up blood investigations were normal. Both cases were treated with oral steroids 1mg/kg body weight and showed significant improvement. Therefore measuring the Homocysteine levels may be a valuable investigation to manage and improve the outcome of patients with retinal vasculitis.

Keywords: Retinal Vasculitis; Vasoproliferative Tumour; Hyperhomocysteinemia; Retinal Tumour; Ocular Inflammation.

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INTRODUCTION

Case 1

31 year old male came with c/o floaters since 15 days with nil systemic history. Both eye BCVA was 6/6. Anterior Segment was normal. RE fundus was normal. LE media was clear. Patient had hyperemic disc, splinter haemorrhage, well circumscribed elevated lesion with tortuous blood vessels over it with surrounding sub retinal exudation and inferior vitreous haemorrhage was noted.

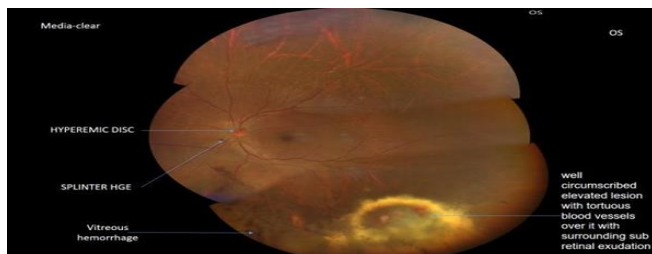


Figure 1: LE fundus showing hyperemic disc, splinter haemorrhage, well circumscribed elevated lesion with tortuous blood vessels over it with surrounding sub retinal exudation and inferior vitreous haemorrhage.



Figure 2: RE Fundus appears normal RE OCT was normal.

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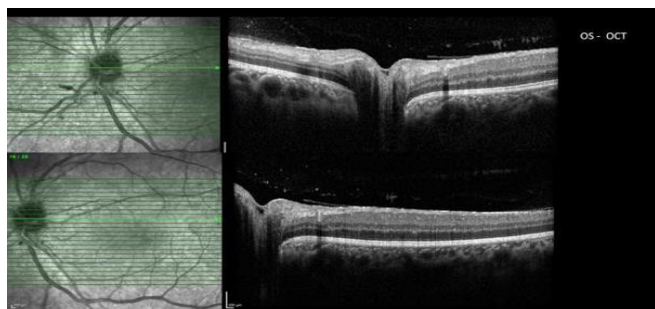


Figure 3: LE OCT showed disc edema with vitreous cells in the inferior vitreous face and thin epiretinal membrane

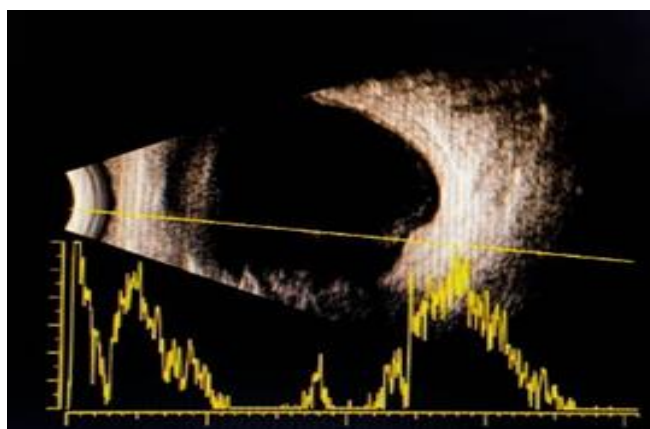


Figure 4: (OS) B scan showed moderately echogenic well circumscribed homogenous mass noted in ITQ corresponding to the clinically seen lesion

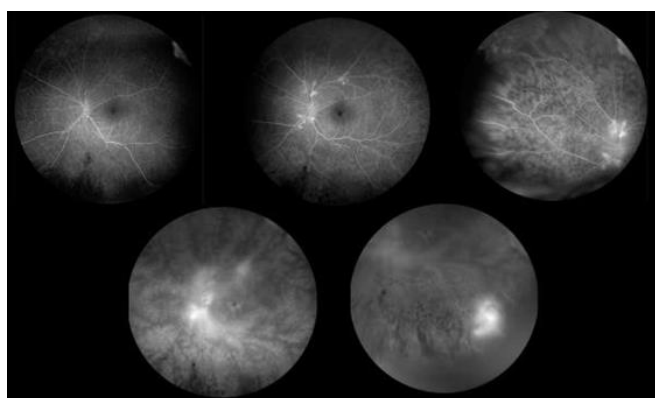


Figure 5: LE FFA showed disc leakage in the AV phase. Capillary ferning in all quadrants in the venous phase. ITQ hyperfluorescent lesion corresponding to the vascular leakage from the VPT was seen. Blocked fluorescence due to inferior vitreous hemorrhage was noted

Blood investigations were done. CBC, ESR, CRP, RBS was normal. CXR and Mantoux was normal. TPHA, HBsAG, HIV and HCV was non-reactive. HLAB51 was negative. Hence we came to the diagnosis of LE Vasoproliferative Tumor Associated With Retinal Vasculitis & Hyperhomocysteinemia.

As serum Homocysteine was elevated patient was referred to the cardiologist & was started on vitamin supplements. As patient had LE retinal vasculitis with VPT Patient was started on Oral Steroids 1mg/Kg Body Weight

CASE 2

A 22 years male. Complaints of gradual painless diminution of vision in RE past 7 years Patient was diagnosed with LE pars planitis elsewhere. Patient is a k/c/o Hypertension x 2 months on treatment and Hyperhomocysteinemia x 2months on treatment. Patient is a known alcoholic & a chronic smoker.

Patient RE exotropia of 15 degree with BCVA 6/36 and LE 6/12. He had circumcorneal congestion in both eyes. RE occasional cells and LE cells 2+ with flare 1+. LE intraocular pressure was 30mmHg.

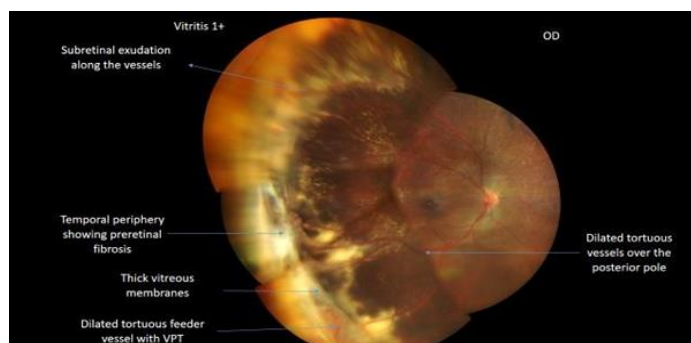


Figure 6: RE fundus showed vitritis 1+,dilated tortuous vessels over the posterior pole,temporal periphery showed pre retinal fibrosis and thick membranes, dilated tortuous feeder vessels with vasoproliferative tumour was noted in the inferotemporal quadrant and subretinal exudation along vessels were noted.



Figure 7: LE fundus showed peripheral sclerosed vessel



Figure 8: LE FFA showed signs of vasculitis with capillary ferning in all quadrants in the late venous phase due to blood retinal barrier impairment.

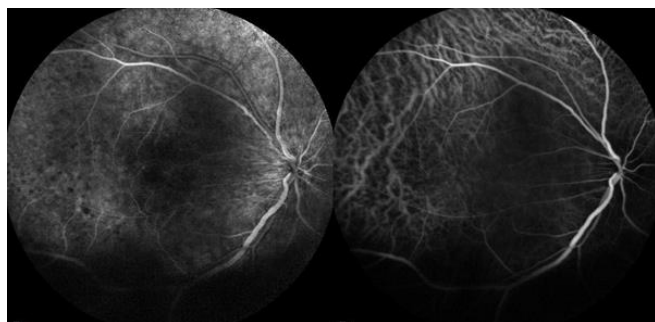


Figure 9: RE FFA also showed capillary ferning in all quadrants in late venous phase with staining of pre retinal fibrosis.

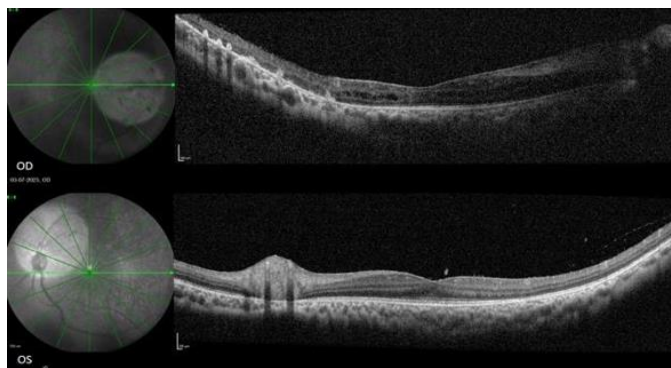


Figure 10: RE OCT showed RPE degeneration in the temporal mid periphery with macular edema. LE OCT showed normal foveal contour

Blood investigations showed elevated Hb 17.8 g/, elevated triglycerides-433 mg/dl and elevated serum homocysteine level: 39.6 micro mol/l. ESR, CRP was normal, vitamin B12 was normal, HLAB51 was negative, mantoux was negative, HRCT chest was normal and RA factor was negative. TPHA, HBsAG, HIV and HCV was non reactive.

Hence we came to the diagnosis of RE Vasoproliferative Tumor With BE Retinal Vasculitis associated with Hypercoagulability, Hyperhomocystenaemia And Polycythemia Vera patient was started on antiglaucoma medication in view of LE high IOP, topical steroids & NSAID.

Patient was referred to cardiologist. Carotid doppler showed no significant abnormality. ECHO Showed Normal LV Systolic & Diastolic Function (LVEF-64%) and was started on T.Ecosprin 75 mg & T.Rozavel 5mg. For peripheral VPT & active vasculitis. Patient was started on topical NSAID and oral steroids 1mg/kg body weight. Post 1 month follow up patient's BCVA improved to RE 6/24 LE 6/9.

There was significant reduction in the subretinal exudates in the temporal periphery and the calibre of the dilated vessels over the posterior pole had reduced.

CASE 3:

A similar case 40 year-old Female was diagnosed with LE intermediate uveitis + peripheral mass ITQ near the pars plana with vitritis 1+ with vision LE 6/9 N6 and RE 6/6 N6. A yellowish mass raising from a snow bank surrounded by vitreous inflammatory snow balls were noted. Tortuous fine retinal vessels within the lesion were evident. RE anterior

segment and fundus was normal.



Figure 11: Hyperemic disc with a yellowish mass raising from a snow bank surrounded by exudates in the inferotemporal quadrant with inferior vitreous haemorrhage.

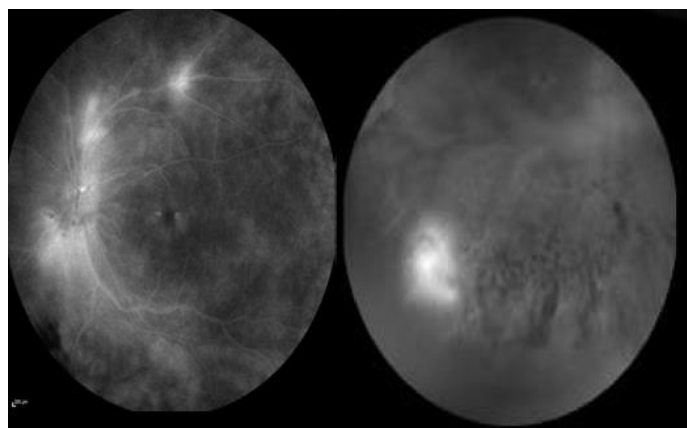


Figure 12: Fundus fluorescent angiography showed signs of vasculitis associated with an hyperfluorescent mass in the peripheral inferotemporal quadrant and blocked fluorescence noted due to vitreous haemorrhage.

Blood investigations was done. CBC, ESR, CRP, RBS was normal. CXR and Mantoux was normal. TPHA, HBsAG, HIV and HCV was non reactive. HLAB51 was negative. Here again Serum Homocysteine level was found elevated (40.6 micro mol/l).

Hence we came to the diagnosis of le vasoproliferative tumor associated with retinal vasculitis & hyperhomocystenaemia. As patient had LE retinal vasculitis with VPT patient was started on Oral Steroids 1mg/Kg Body Weight.

In this case too as serum Homocysteine was elevated hence patient was referred to the cardiologist & was started on vitamin supplements.

DISCUSSION

VPT appear as a globular- or dome-shaped lesion arising in the peripheral retina. It is yellow or pink in colour, occasionally -white, grey, or reddish in appearance.

The vascular nature of the lesion manifest as a network of fine, telangiectatic vessels over the surface of the lesion. Clusters of small aneurysms of varying size are present within the telangiectatic plexus. Retinal 'feeder' vessels have a normal / slightly enlarged caliber enter the posterior aspect of the tumour. The microscopic study shows that VPT is a mix of both glial and vascular component. Glial component exhibits fusiform cells. Vascular component show dilated vessels with mural hyalinization and occasional thrombosis the proliferative index of vpt is low.

Walinjkar JA studied 22 tumors from 19 eyes of 17 patients with mean age at presentation- 43.5 years (range: 15–68 years), mean (BCVA) was + 1.10 logMAR out of which 68% eyes had secondary tumor and 10% tumors involved IQ.

The studied showed that most common association of secondary VPT were Coats disease, retinal vasculitis, PCV and FEVR.

Tumor-associated features were Intra/subretinal exudates, Vitritis, SRF, VH preretinal fibrosis, ERM and SRH.

The treatment given to these patients with VPT were cryotherapy, intravitreal /oral steroids, laser photocoagulation, anti-VEGF and few were under observation.

Angad Miskin et al conducted a case control study to investigate relation between Serum Homocysteine Levels and Retinal Vasculitis at tertiary care centre in North India.

The study concluded that there is no significant association between raised serum homocysteine levels and retinal vasculitis.

Soha Senara et al wanted to find out the possible relation between serum homocysteine level and ocular involvement in Bechet's patient.

Patients were divided in 2 groups those with Bechet's disease and eye involvement and those without ocular involvement. Homocysteine readings were higher in BD patients as compared to normal control group.

Measured the HCY levels in extraocular Bechet's disease and ocular Bechet's disease found there was no statistically significant difference between the two. However the study

concluded that there was no association between homocysteine level and eye involvement.

Jyothirmay Biswas et al concluded a prospective study to investigate the increased homocysteine levels and its metabolite thiolactone in patient with Eale's disease. The study showed that increased homocysteine level and its metabolite thiolactone increased the oxidative stress in patient in Eale's disease.

CONCLUSION

Hyperhomocysteinemia is a treatable risk factor. Measurement and monitoring of Homocysteine levels may be a valuable index in the investigation, management and improving the outcome of patients with retinal vasculitis.

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