Retinal tumors are a rarity with peripheral retinal tumors being still more rare. Here we report a case where a peripheral retinal tumor presented with a progressive decrease in vision over a long time due to cystoid macular edema.

A 37 yr old male presented with history of gradual, painless progressive loss of vision left eye since 2 years, with no improvement with treatment. On examination, he had a best corrected visual acuity of 6/6 in the right eye and 6/18 in left eye. Intraocular pressure in both eyes was normal. Examination of the right eye including anterior and posterior segments was within normal limits. Anterior segment examination of the left eye revealed 1+ cells and haze in the anterior chamber. Vitreous showed 3+ cells and 2+ haze with membranes and strands signifying partial PVD. Fundus examination revealed hyperemic disc and stereoscopic examination of the macula revealed the presence of cystoid macular edema. There was fibrous proliferation and epiretinal membrane along the arcades. A yellowish pink elevated mass about 7 to 8 DD in size, with irregular surface was seen superotemporally beyond the equator. Vessels traversing and supplying the mass appeared dilated and mildly tortuous and there were retinal pigment epithelial changes at the base of the mass all around. There were some telangiectatic vessels over the mass along with hemorrhage. The lesion was also associated with some intraretinal and subretinal exudation (Fig 1).

Fluorescein angiography showed patchy hyperfluorescence of lesion with multiple areas of window defects corresponding to the RPE changes and staining and blocked fluorescence secondary to the exudation and mass lesion. The hyperfluorescence increased in the late phase due to leakage in some areas. There were areas of blocked fluorescence over the lesion corresponding to areas of hemorrhage and in the periphery of the lesion secondary to the RPE hyperplasia. There was late disc leakage and flower petal leakage at fovea signifying cystoid macular edema (Fig 2).

OCT showed macular edema with a cystoid pattern with a central macular thickness of about 450 microns.

B Scan showed an elevated, acoustically solid mass about 9.7 x 7.1 x 3.2 mm in size and multiple echoes in the vitreous suggestive of opacities (Fig 3).

The following differential diagnosis were entertained

1 Vasoproliferative tumour
2 An inflammatory or infective mass
3 Angioma of Von Hippel Lindau disease
4 Coats disease

A complete systemic evaluation was sought to rule out coexistent systemic infections. Mantoux test was found to be negative after 48 hours. Peripheral smear showed normocytic normochromic blood picture. USG abdomen, CT Thorax and MRI brain (plain and contrast) were
normal. Therefore infective or inflammatory mass was ruled out. However, cholesterol levels were found to be high (496.4mg/dl).

During the differential diagnosis, Von Hippel-Lindau was excluded because of absence of grossly tortuous and engorged blood vessels, absence of family history for the disease and absence of other features of Von Hippel-Lindau disease. In VHL even small tumours are associated with grossly engorged and tortuous blood vessels. Also the angiographic characteristics of Von Hippel-Lindau like rapid filling of arteries and rapid AV transit were not seen.

Coats disease was excluded because that is usually seen in boys at a younger age, and usually does not cause tumour like lesions even when exudation is severe. The exudations in Coats disease are often flat and not elevated. Besides tumour like lesions, if present in Coats are accompanied by advanced exudative retinal detachment.

Our patient showed the typical clinical picture of vasoproliferative tumour. He had a solitary, unilateral, yellowish vascularized tumour associated with intraretinal exudation, RPE hyperplasia at the base, telangiectasias and hemorrhage over lesion. He had additional findings of anterior chamber cells, vitreous cells and cystoid macular edema all of which have been reported in various literature 1,2,3. We believe these changes to be a secondary reactionary process to the presence of tumour in the eye. FFA picture further helped to confirm the diagnosis. Presence of elevated levels of cholesterol in patients with vasoproliferative tumour has been reported in literature 4.

With the diagnosis of vasoproliferative tumour, decision to treat with multiple sessions of cryotherapy was taken and patient given first session of cryotherapy. He is at present on follow up.

**Discussion**

The term vasoproliferative tumour was coined by Shields et al in 1995. Previously these tumours were called presumed acquired haemangiomas, angioma like lesions and peripheral retinal telangiectasia.

Vasoproliferative tumours of the retina are benign vascular tumours of unknown origin. These tumours generally present as yellow pink, one or more retinal nodules seen usually in the pre-equatorial fundus, generally in the inferotemporal quadrant but may also be seen in the upper retinal quadrants 2,5 or even at posterior pole 6,7. Their feeding and draining vessels are slightly dilated but not enlarged or convoluted. These tumours can be associated with additional clinical changes like intraretinal and subretinal hemorrhages, intraretinal and subretinal exudation, exudative retinal detachments, hyperpigmentation of RPE, vitreous and anterior chamber cells, vitreous hemorrhage, preretinal macular fibrosis and macular edema 1,2,7. Exudation in vasoproliferative tumours tends to creep back towards the fovea and is hence seen in continuity with the lesion 4,7. However preretinal gliosis can occur remote from the lesion 7.

The pathogenesis of these tumours has not been established. In a study of 103 patients, Shields et al found that these lesions were idiopathic or primary in 74 % and secondary to congenital, inflammatory, vascular, traumatic, dystrophic and degenerative ocular diseases in the rest 1. Primary vasoproliferative tumours are solitary, unilateral and generally located in the inferotemporal quadrant of the retina. Many of the patients with primary vasoproliferative tumours also have systemic hypertension. Secondary vasoproliferative tumours on the other hand are bilateral, multifocal and can be located in any quadrant of the retina. Secondary
tumours also tend to be more ill defined and diffuse \(^7\).

These tumours are believed to represent gliovascular proliferations with varying degrees of both gliosis and vascular proliferation. Histopathology of these tumours shows them to be composed predominantly of elongated, spindle shaped cells, corresponding to glial cell origin imposed over a fine capillary background. Mitotic figures, pleomorphism or cellular atypia has not been shown to be present. Another important feature of these tumours is the presence of dilated blood vessels within the tumour mass \(^2,5\).

Various treatment options have been described for vasoproliferative tumours like periodic observation \(^1,2\), cryotherapy \(^1,2,3,4\), laser photocoagulation \(^1,4\) and plaque radiotherapy \(^1,2,8\). Some reports suggest that these tumours may be treated successfully with photodynamic therapy. Vitrectomy may also be needed for complications accompanying these tumours like vitreous haemorrhage and retinal detachment. Irvine F et al\(^9\) have suggested that trans scleral resection be attempted if there is difficulty in diagnosis. This would give tissue for diagnosis and also avoid unnecessary enucleation in case of diagnostic dilemma.

In conclusion, vasoproliferative tumours should be included in the differential diagnosis of any peripheral retinal tumour. They should be recognized by their distinctive clinical features and angiographic characteristics and being decidedly benign their recognition would avoid unnecessary morbidity for the patient.

### References