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Pseudoangiomatous retinal gliosis (PARG) treated with iodine plaque in patient with chronic retinal detachment

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ABSTRACT

Purpose: To describe a case of a chronic retinal detachment complicated by the development of pre and subretinal hemorrhage secondary to a large pseudoangiomatous retinal gliosis (PARG) that interfered with retinal reattachment. After the lesion was regressed following plaque radiotherapy retinal reattachment was successfully completed.

Observations: A 56y.o healthy man with known history of a chronic inferior rhegmatogenous retinal detachment (RD) of the left eye (OS) presented to the Bascom Palmer Eye Institute (BPEI) emergency department (ED) complaining of new floaters OS. On examination, the patient had a visual acuity of 20/30 right eye (OD) and 20/200 OS. Fundoscopic examination showed a treated tear in OD and dense vitreous hemorrhage OS. Initial B-scan ultrasonography OS showed an inferior RD with diffuse hyperechoic material in the vitreous cavity, preretinal and subretinal space most consistent with hemorrhage. Three days later the patient presented with further vision loss and a repeat B scan showed total RD and increasing subretinal hemorrhage with a solid mass like lesion. At this point, decision was made to proceed with retinal detachment repair, removal of the vitreous hemorrhage, and retina evaluation. During surgery, a total retinal detachment was encountered with poor view of the inferior retina due to a large round vascular lesion in the subretinal space with surrounding hemorrhage and clots. The retina was reattached during surgery, however, the postop was complicated by recurrence of VH, dense hyphema, increased IOP, recurrence of retinal detachment, and growth of the mass like lesion noted during surgery. Consultation with ocular oncology diagnosed the patient with secondary PARG lesion and plaque radiotherapy was given achieving remarkable regression of the lesion. After the lesion had regressed, successful retinal reattachment was achieved, and the patient had excellent visual recovery.

Conclusion and importance: PARG lesions are uncommon in particular when associated to chronic retinal detachments. This case highlights the importance of having a high clinical suspicion for the development of these lesions to diagnose them correctly and treat them aggressively with plaque radiotherapy in order to be able to manage the underlying complex retinal detachment.

1. Introduction

Pseudoangiomatous retinal gliosis (PARG) also known as presumed acquired retinal hemangioma,[1] angiomatous or angioma-like retinal tumor,[2–4] vasoproliferative retinal tumor,[5–12] reactive retinal glioangiosis,[8,13] and reactive retinal astrocytic tumor[14] usually presents as a solitary and unilateral lesion with predilection for the inferotemporal peripheral retina and most commonly affects the older population. Typical features of this lesion include a circumscribed grayish-pink retinal mass, subretinal lipid exudation, focal intralesional hemorrhages, and exudative retinal detachment.[9] It resembles the retinal capillary hemangioma (RCH) but can be distinguished from it due to the lack of association with Von Hippel-Lindau syndrome (VHL), no family history, older age range of affected individuals, and for the ophthalmological findings which include the absence of the typical dilated feeder artery and draining vein seen in RCH.[1] It is often associated with an underlying inciting chorioretinal disorder.[5] Ultrasonography typically demonstrates medium to high internal reflectivity on the A-scan and acoustic solidity without choroidal excavation.[5] Histopathologic examination is characterized by the presence of predominantly glial cells.[14] Differential diagnoses include retinal capillary hemangioblastoma, peripheral exudative hemorrhagic chorioretinopathy

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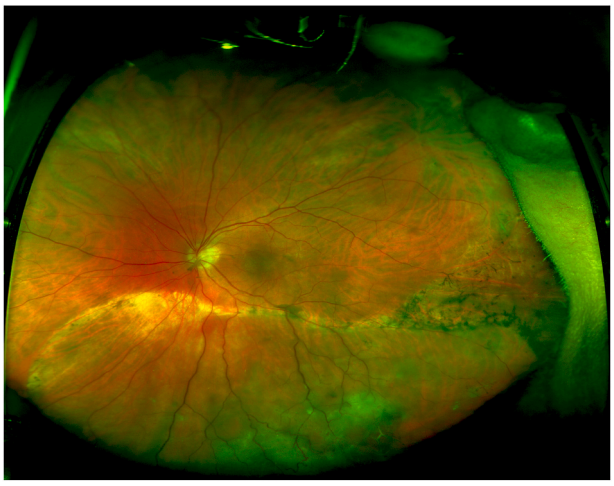

Fig. 1. Fundus image of OS showing inferior chronic RD with demarcation line at initial presentation 2 years prior to RD.

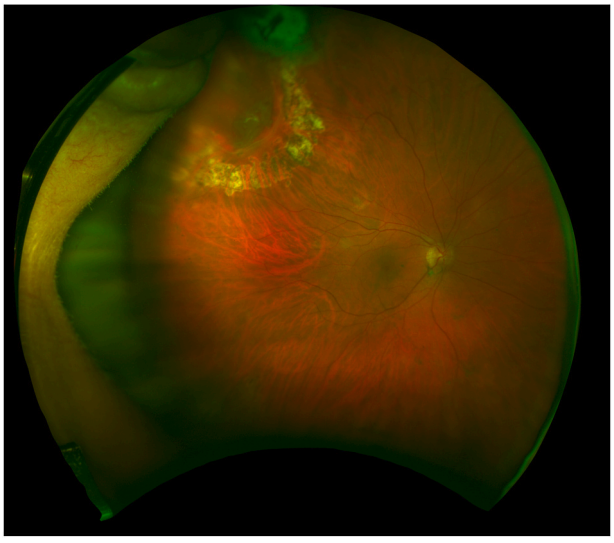

Fig. 2. Fundus image of OD at initial presentation showing well treated retinal tear.

(PEHCR), choroidal melanoma, RPE adenoma/adenocarcinoma, and Coat's disease.[1]

We report a case of a patient with a known history of a chronic inferior rhegmatogenous retinal detachment (RD) and proliferative vitreoretinopathy who was lost to follow up and presented with subretinal, pre-retinal, and vitreous hemorrhage secondary to a large PARG lesion. The PARG lesion was successfully treated with a radioactive plaque which allowed a clear view for adequate management of the chronic retinal detachment. The patient had successful repair with excellent visual recovery despite the complex course and the recurrent retinal detachments.

2. Case report

A 56y.o Caucasian male with no significant past medical history presented to clinic with a four-day history of flashes and floaters in his OD. On ophthalmic examination best corrected visual acuity (BCVA) was 20/20 OD and 20/40 OS. Schaffer sign was present in the OD. Fundoscopy OD revealed vitreous hemorrhage and multiple areas of lattice with atrophic holes as well as an 11 O'clock horseshoe tear associated with subretinal fluid. Incidentally, examination of the OS revealed a chronic inferior rhegmatogenous RD with demarcation line and fluid at level of inferior arcade that extended from 0400 to 0800 O'clock secondary to retinal holes in lattice inferotemporally. The OS showed peripheral membranes due to proliferative vitreoretinopathy (PVR) and a macular epiretinal membrane (ERM) (Fig. 1).

Laser retinopexy was performed to his OD and decision was made to follow OS closely until OD acute lesion was resolved before considering retinal detachment repair with membrane peel of the left eye. One month after retinopexy, VA improved to 20/25 OD with a well demarcated retinal tear (Fig. 2). At this visit, the patient was offered surgical repair of OS, but the patient decided to delay surgery and schedule a follow up. However, the patient was lost to follow up.

Two years later the patient presented to the emergency department complaining of new floaters in his OS, BCVA was 20/30 OD and 20/200 OS, on fundus examination OS he had new vitreous hemorrhage. Bedside B-scan showed vitreous hemorrhage with the inferior RD seen on previous exam with posterior retina attached. The patient was scheduled for follow 3 days later with possible surgical repair within 1 week. However, 2 days later the patient presented due to new inferior visual field defect and B scan revealed bullous extension of the RD superiorly with possible temporal tears and a mass like lesion inferiorly with diffuse subretinal hemorrhage (Fig. 3). Retinal detachment repair and retina evaluation were scheduled for management of the complex total retinal detachment with large hemorrhagic component. The patient was not on any blood thinners.

Surgery was performed with placement of a scleral buckle prior to the vitrectomy. During the vitrectomy, the retina was found to be totally detached due to a combined rhegmatogenous and exudative retinal detachment, with the superior retina bullously detached from 9 to 3 in a clockwise fashion and the inferior retina was also detached from 3 to 9 with a large preretinal hemorrhagic clot and a large subretinal hemorrhagic clot. Within the area of inferior retinal detachment, the preretinal, intraretinal and subretinal hemorrhage precluded complete evaluation of the tissue and did not allow to completely rule out an underlying retinal or choroidal lesion or mass. Nasal and temporal tears were noted and lasered. Inferior-temporal holes over the lattice were lasered. Additionally, 360 laser was performed except over inferior retina where preretinal hemorrhage precluded adequate laser uptake. Gas tamponade was used, and vitreous fluid was sent to the pathology department, which came back negative for malignant cells.

Post-operative retina evaluation was limited due to post-op vitreous hemorrhage. One week after surgery the patient presented with orbital pain due to a large hyphema with intraocular pressure (IOP) of 40 mmHg. B-scan revealed attached retina with superior gas bubble and inferior retinal lesion with high internal reflectivity and UBM showed hyphema, without any masses in iris or ciliary body. IOP was improved with medical treatment and complete resolution of hyphema was achieved. Two weeks after surgery, a follow up B scan showed peripheral

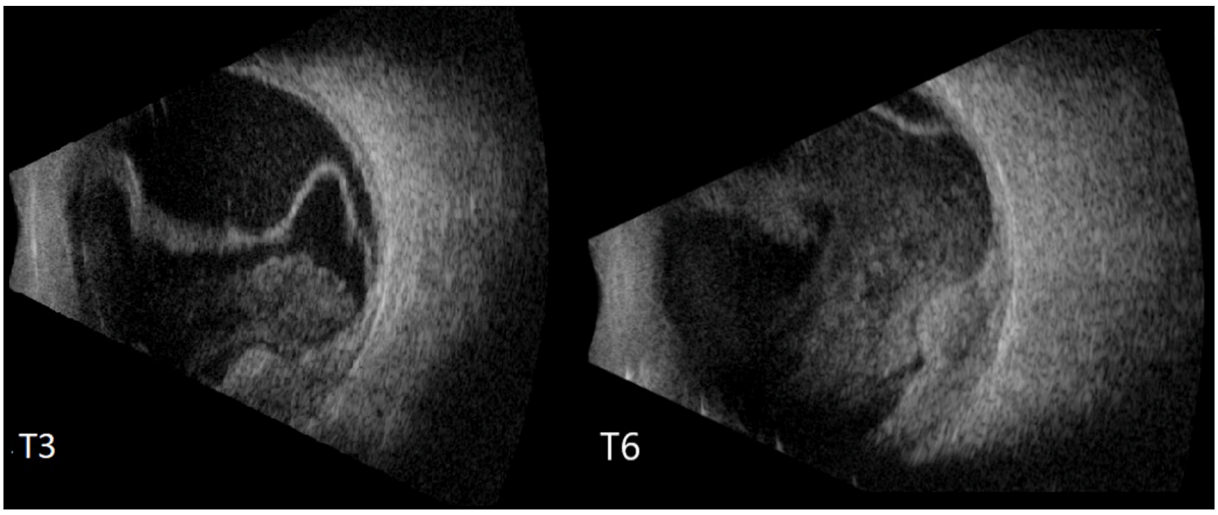

Fig. 3. Temporal (T3) and inferior (T6) transverse Bscan of left eye. Both images were done prior to initial RD repair showing bullous superior RD, shallow inferior RD with mass like inferotemporal lesions showing diffuse hyperechoic material in the vitreous cavity, preretinal and subretinal space consistent with hemorrhage.

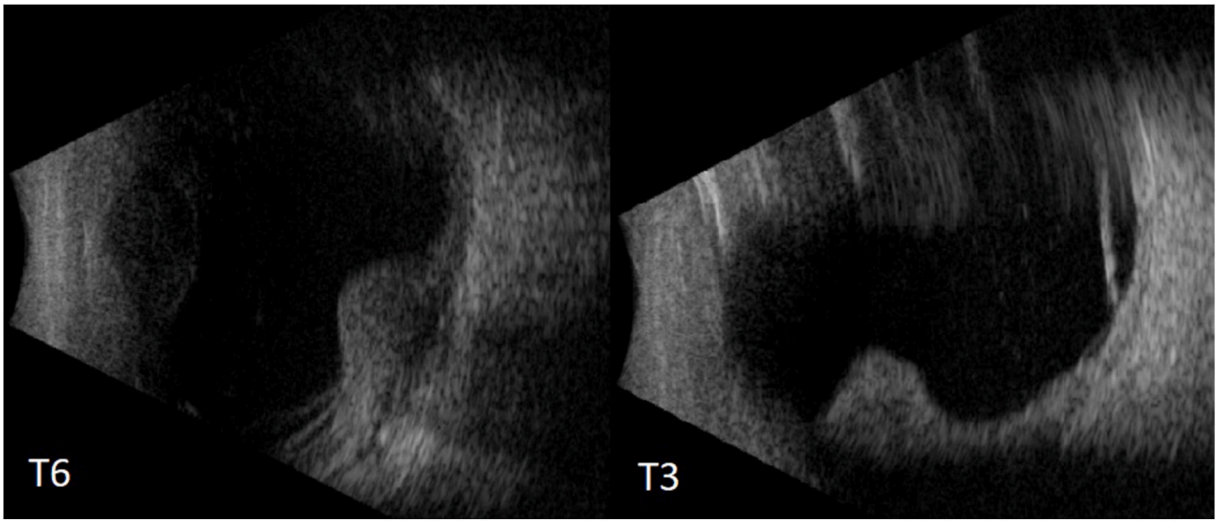

Fig. 4. T6 and T3 transverse Bscan showing enlarging hyperechoic retinal lesion with early recurrence of RD and residual superior gas bubble.

nasal shallow RD under the residual gas bubble and an enlarging inferior solid lesion of 14.10 mm × 6.27mm (Fig. 4). Given the growth of the lesion, ocular oncology was consulted.

Their evaluation concluded that the lesion represented a large PARG secondary to the prior chronic retinal detachment and the decision was made to proceed with plaque radiotherapy with the goal of regressing the lesion to allow subsequent reattachment of the retina. Radioactive plaque with a therapeutic dose of 30 Gy delivered at 7.3 mm depth was placed for three days. Fundus exam repeated 10 days after plaque radiotherapy showed remarkable resolution of the hemorrhaging with improved retinal view and marked regression of the vascular lesion (Fig. 5). Three weeks after the lesion was plaqued near complete regression was noted and RD repair with cataract extraction and IOL placement was scheduled. In this surgery, 360 recurrent RD was noted, and the inferior lesion showed signs of complete regression, retina was reattached, and silicone oil tamponade was placed. Postoperatively the retina remained attached without recurrence of the tumor. Silicone oil was removed and an epiretinal membrane was peeled off the surface of the macula. At the most recent follow-up, which was 4 years after the plaque and RD surgery, his vision in OS had recovered to 20/40 with

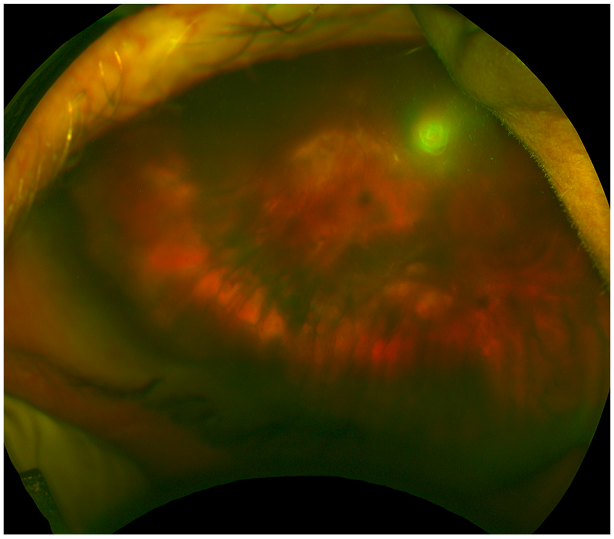

Fig. 5. Fundus image 10 days after plaque insertion showing lesions regression and improvement of vitreous hemorrhage.

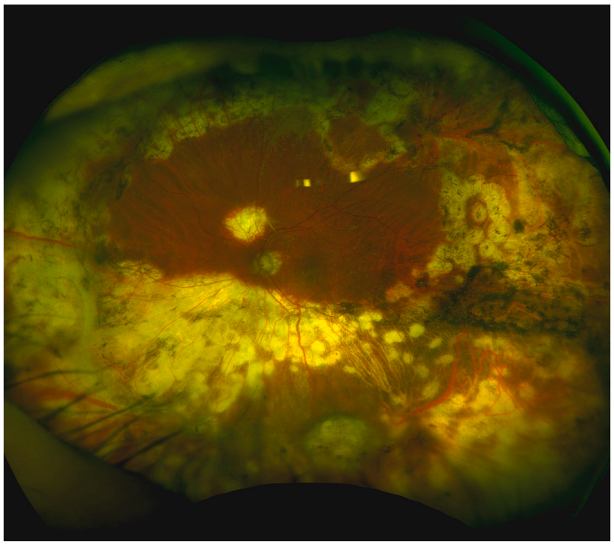

Fig. 6. Fundus image 4 years after plaque and RD repair, showing regressed and fibrosed lesion inferiorly and retina attached with prior retinal scars.

best correction and on fundoscopic examination, the retina was nicely attached with complete regression of the PARG lesion (Fig. 6) and B scan images showed regressed lesion post RD repair (Fig. 7).

3. Discussion/conclusions

PARG represents a well-defined clinical entity that can readily be distinguished from simulating conditions such as true RCH and adult Coats' disease. PARG is frequently associated with abundant exudation causing retinal detachment and can be associated with vitreous hemorrhage,[5] it's important to remember that this pathology can develop secondary to a preexisting ocular disease.[1,3–5,12] Treatment varies from observation to laser or plaque radiotherapy and although there is still not a clear path as to how to treat this lesion, the choice of treatment can be based on tumor size, location, and associated vitreoretinal findings.

In our report, the patient had a previous diagnosis of chronic rhegmatogenous RD, which is one of the chorioretinal lesions seen in cases that develop PARG. The fact that we saw the patient 2 years before this presentation and that we had previous fundus images helped us narrow the differential diagnosis. Due to the previous history of chronic RD, no previous lesion/mass located at last exam of the patient, no other medical problems, negative malignant cells on vitreous fluid sent to pathology, characteristic appearance on the US (medium to high irregular internal reflectivity with no choroidal excavation) and the characteristic appearance and location of the lesion on exam (circumscribed, peripheral, nodular and of yellowish-red color), we were able to make a presumed diagnosis of PARG with a combined exudative and rhegmatogenous retinal detachment.

In a case series by Cohen et al.[15] of 30 patients with PARG lesions, showed that the use of plaque radiotherapy can induce rapid regression of the lesions. Based on the size of the lesion (>2.5 mm thickness) and the location of the lesion in our case, it was treated with plaque radiotherapy. The lesion showed rapid and remarkable regression of the vascular abnormality and associated hemorrhaging allowing us to successfully manage the rhegmatogenous component of his retinal detachment.

Despite the benign histopathologic features[8,9] and peripheral location of PARG lesions, vision loss can occur secondary to associated vitreoretinal findings, including cystoid macular edema, macular exudation, subretinal fluid, preretinal fibrosis, and vitreous hemorrhage.[1,5] Total exudative retinal detachment with secondary glaucoma leading to eventual enucleation[16] has been previously reported.

PARG lesions are uncommon in particular when related to a retinal detachment, thus it is important to have a high suspicion in order to identify these lesions. A multispecialty approach to these cases including the use of plaque radiotherapy can help regress the lesions to allow for successful retinal detachment repair and to achieve a successful clinical outcome as in this case.

Patient consent

Consent to publish this case report was not obtained. The report does not contain any personal information that could lead to the identification of the patient.

Authorship

All authors attest that they meet the current ICJME criteria for Authorship.

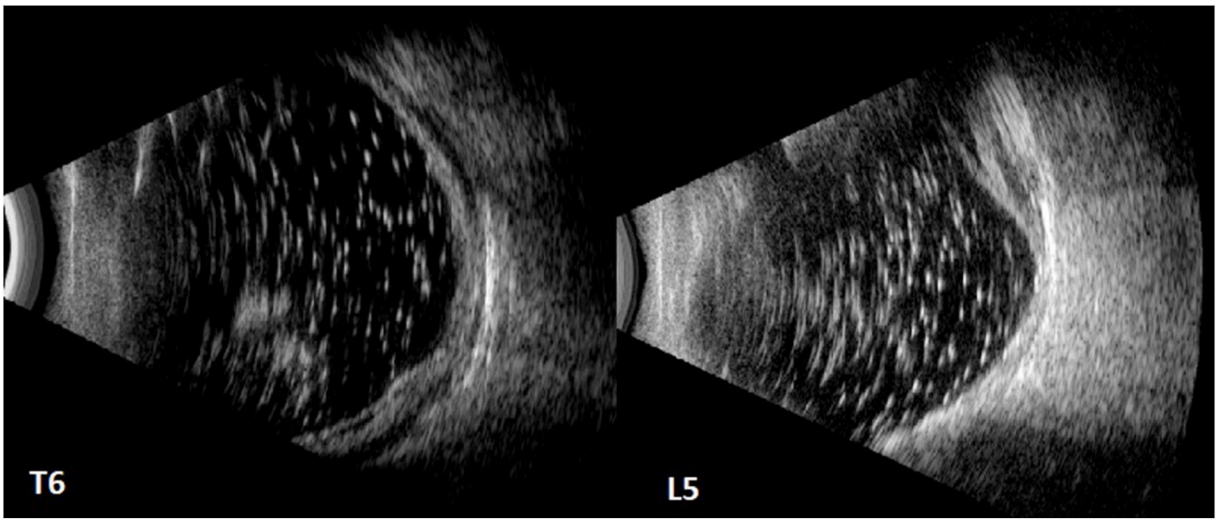

Fig. 7. T6 (transverse) and L5 (longitudinal) Bscan 4 years after plaque and RD repair, showing decreased size of hyperechoic retinal lesion and presence of hyperechoic particles in the vitreous cavity consistent with residual silicone oil droplets.

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