



Case report

Combined rhegmatogenous and traction detachment associated with vasoproliferative tumor secondary to sickle cell retinopathy



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ABSTRACT

Purpose: To report the surgical management of a combined rhegmatogenous and traction retinal detachment associated with a vasoproliferative tumor secondary to sickle cell retinopathy.

Observations: A 29 year old man from Ghana presented with unilateral vision loss, ischemic retina and sea fan neovascularization in both eyes and a retinal detachment nearby a vasoproliferative tumor (VPT) in the left eye. Hemoglobin electrophoresis led to the diagnosis of sickle cell disease. The patient underwent vitrectomy with scleral buckle surgery, resection of the tumor, and removal of subretinal membranes in the left eye. Laser photocoagulation was targeted to areas of ischemic retina in both eyes. **Conclusions:** and **Importance:** To our knowledge, this is the first report of a combined rhegmatogenous and traction retinal detachment associated with a VPT in sickle cell retinopathy managed by modern vitrectomy techniques. Prompt recognition of the condition and surgical management addressing both rhegmatogenous and tractional components can lead to improved outcome.

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1. Introduction

Vasoproliferative tumors are described as benign masses of neovascularization.¹ Approximately 75% of reported cases are idiopathic or primary VPT occurring mostly in healthy patients.² Secondary VPT have been associated with uveitis,³ retinitis pigmentosa, chronic retinal detachment, Coats' disease, prior retinal detachment repair, idiopathic peripheral retinal vasculitis, familial exudative vitreoretinopathy, toxoplasmosis, aniridia, congenital hypertrophy of retinal pigment epithelium, idiopathic choroiditis, retinopathy of prematurity, and histoplasmosis.² We report the management of a VPT associated with a traction and rhegmatogenous retinal detachment secondary to sickle cell retinopathy (SCR).

2. Case report

A 29-year-old man from Ghana presented with loss of vision in the left eye for several months. Vision was 20/20 in the right eye and count fingers in the left eye. Retinal exam showed ischemic retina and multiple areas of fibrovascular proliferation in each eye

(Fig. 1). Additionally, a vascularized mass with dilated feeder vessels was found in the temporal periphery along with a combined tractional (TRD) and rhegmatogenous retinal detachment (RRD) with a small retinal hole in the left eye. Widefield fluorescein angiography (FA) (Fig. 2) revealed sea-fan neovascularization in the peripheral retina. Ultrasound measurements of 3.6 mm × 16.2 mm × 18.2 mm and high internal reflectivity through the lesion were suggestive of a vascular tumor (Fig. 3). Hemoglobin electrophoresis revealed the presence of sickle cell (SC) trait leading to the diagnosis of SCR.

Due to the retinal detachment, pre-operative anti-VEGF therapy, laser or cryotherapy was deferred. The patient underwent 23-g pars plana vitrectomy with placement of a scleral buckle encircling band to support the vitreous base. Triamcinolone aided identification and removal of the posterior hyaloid. Intra-operatively, the mass was found to cause considerable foreshortening of the retina and reattachment could not be performed without resecting the tumor. Diathermy was used to cauterize the feeder vessels and then to circumscribe the lesion prior to resection of the tumor. Unfortunately, the resected lesion was unable to be adequately recovered by the pathologist for further tissue identification and histopathology. Nearby subretinal membranes were resected through the newly created localized retinectomy. Laser photocoagulation was then applied to the retinectomy as well as areas of ischemic retina.

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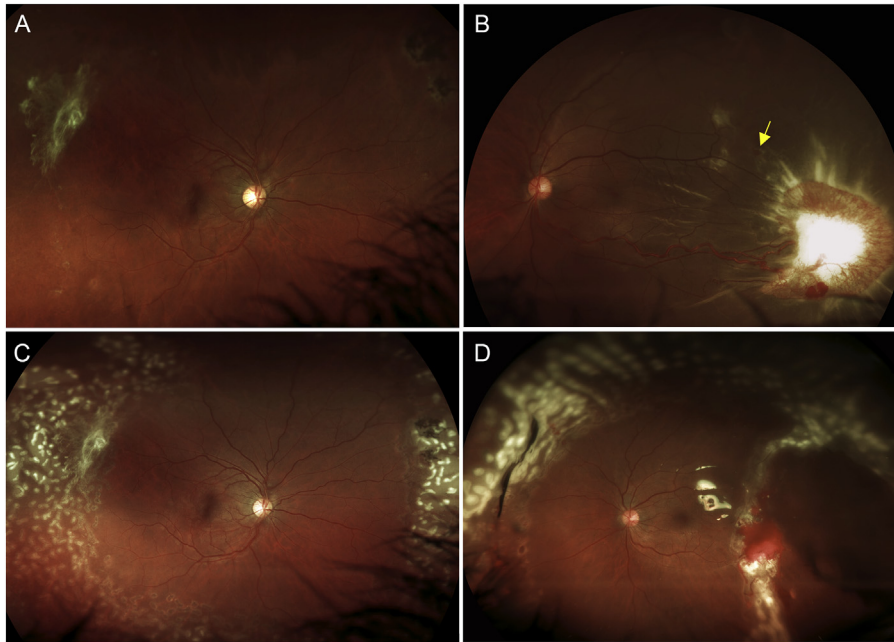


Fig. 1. Color Fundus Photography. (A) Right eye showing infarcted sea-fan neovascularization in the temporal periphery as well as ischemic retina and ghost vessels. A black sunburst is seen in the superonasal retina. (B) Left eye showing dilated vessels and vascularized mass in temporal periphery. Subretinal fibrosis and an atrophic hole (yellow arrow) has led to a combined tractional-rhegmatogenous retinal detachment. Subretinal fluid is seen within the macula and beneath the fovea. (C) Right eye 1 week following laser photocoagulation to ischemic retina. (D) Left eye 1 week following vitrectomy, resection of vasoproliferative tumor and removal of subretinal membrane. Laser photocoagulation is applied to the ischemic retina as well as the retinectomy and along the scleral buckle. Silicone tamponade was placed over an attached retina. (For interpretation of the references to color in this figure legend, the reader is referred to the web version of this article.)

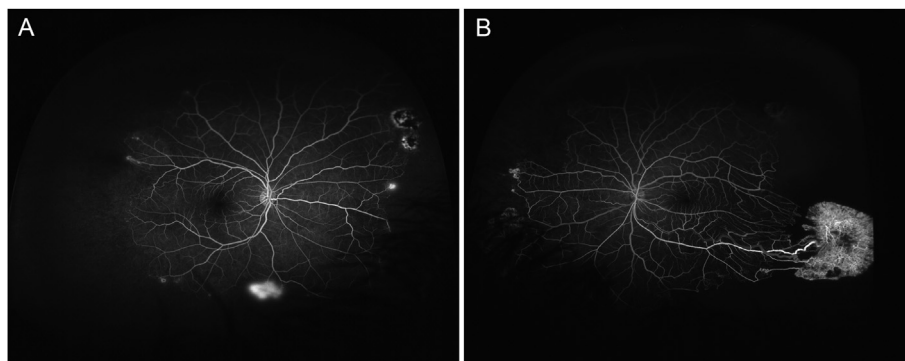


Fig. 2. Wide-field fluorescein angiography. (A) Mid-phase angiogram of the right eye showing capillary non-perfusion in the mid-to far periphery. Neovascularization of the retina as well as a black sunburst lesion (superonasal) are seen. (B) Early-phase angiogram of the left eye showing dilated and tortuous vasculature leading up to a highly vascularized mass lesion. Widespread capillary non-perfusion and neovascularization is also seen.

Silicone oil tamponade was placed. The right eye was also treated with laser photocoagulation to the areas capillary non-perfusion. Post-operatively, the patient developed cataract in the left eye which was later replaced with a posterior chamber intraocular lens. Four months after silicone oil removal, his retina remained attached and his vision improved to 20/400.

The patient provided written consent for publication of personal information including medical record details and photographs.

3. Discussion

Sickle cell disease results from a single point genetic mutation that results in a structural change to the hemoglobin polypeptide chains which disrupts erythrocyte architecture, promoting cellular dehydration and resulting in the sickled appearance of red blood

cells.⁴ The sickled erythrocytes can result in ischemia and infarction of local tissues.⁵ Sickle cell retinopathy (SCR) is found more commonly in the SC and S- β thalassemia subtypes of compound heterozygotes which can be differentiated through hemoglobin electrophoresis.^{6,7} In patients with SCR, occlusion of the retinal vasculature can lead to areas of profound capillary non-perfusion where neovascular lesions can develop and progressively increase in number and size often developing into shapes of sea fans.⁶ Though spontaneous regression, or autoinfarction, of the retinal neovascularization can occur, sickle cell retinopathy is strongly associated with vision loss in as many as 10% of affected patients primarily through the development of vitreous hemorrhage or retinal detachment.⁶

Retinal neovascularization has been associated with retinal vascular diseases including diabetic retinopathy, retinal vein

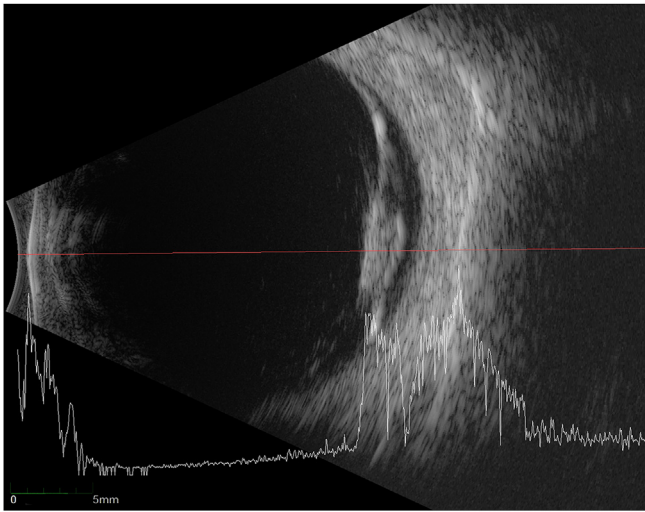


Fig. 3. A and B Scan ultrasonography through the lesion on the left eye. A-scan measurements show high internal reflectivity suggestive of a vascularized lesion. Low reflectivity beneath and surrounding the lesion is consistent with a retinal detachment.

occlusions, retinopathy of prematurity and sickle cell retinopathy.⁸ A strong VEGF presence, associated with retinal and subretinal neovascularization, has been found in both neovascularized retinas of sickle cell patients and in surgically removed VPT.^{1 6 9} Furthermore, vasoproliferative tumors showed regression with anti-VEGF therapy in two different reports.^{1 3} Given these reports, there may be biological plausibility that amount of ischemia seen in patients with SCR may be enough to stimulate the growth of a secondary VPT through a VEGF mediated pathway.

The first association of a VPT with sickle cell disease was made by Galinos et al. in 1979 where they reported a 52 year old Haitian man with SC disease who presented with bilateral angioma-like lesions with overlying fibrous membranes.⁹ The patient underwent scleral buckle surgery, external subretinal fluid drainage, and diathermy to destroy the tumors. Unexpectedly, the patient died six days following surgery. Histological examination of the eye showed sickled erythrocytes in the retinal neovascular proliferations within a matrix of PAS positive material. Pathological reports of VPT from other causes show significant glial cell proliferation along the vascular tumors.¹⁰ Machemer suggested that these fibrous membranes might be the result of leakage of proteinaceous exudate from abnormal vasculature.¹¹ Alterations in the overlying vitreous caused by leaking retinal vascular tumors may result in vitreous separation leading to breaks in the internal limiting membrane through which glial cells can migrate and proliferate.¹² These membranes can lead to traction of the retina and in some cases lead to macular epiretinal membrane or peripheral traction detachment.¹³ McDonald and colleagues reported ten cases of traction macular detachment in patients with peripheral retinal angioma and in five of them, additional significant peripheral traction detachments were observed.¹³ In our case, it is conceivable that with peripheral traction, nearby atrophic hole formation occurred leading to the rhegmatogenous component of the detachment.

When central vision is threatened by macular edema or serous detachment, treatment for VPT can be performed using laser photocoagulation, cryotherapy, or both and may result in tumor regression.¹⁴ More recently, anti-VEGF therapy has been shown to be helpful as an adjunctive treatment.¹³ Vitrectomy surgery with specific attention to fibrous components of VPT is generally reserved for visually significant epiretinal membrane (ERM) formation, vitreous hemorrhage, TRD, RRD, and proliferative vitreoretinopathy (PVR) and can lead to good results.¹³ Tumors can be treated prior to or during vitrectomy surgery. Pretreatment with cryotherapy or laser to the tumor in uncomplicated cases (e.g., ERM) may be helpful.¹³ In eyes where significant traction is present, as in cases of TRD or PVR, pretreatment may lead to worsening traction and should be performed with caution.¹³ In patients with sickle cell retinopathy, previous reports suggested that placement of a scleral buckle can lead to post-operative ocular ischemia; however, a more recent report have shown this risk to be minimal and scleral buckling should be considered if deemed beneficial.¹⁵

Clinicians should be aware of the possible association of VPT with SCR. Serum hemoglobin electrophoresis can be helpful in diagnosing the underlying hematologic condition. Prompt recognition and surgical management of the potential complications associated with VPT and SCR when appropriate can lead to improved outcomes.

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