Case Report

# Vasoproliferative Tumor Secondary to Sarcoidosis-Associated Intermediate Uveitis

Sara Abdel Jalil, Tareq Jaouni, Radgonde Amer

Hadassah Medical Center, Department of Ophthalmology, Jerusalem, Israel

## Abstract

We report the visual and clinical outcomes of a middle-aged woman who presented with exudative retinal detachment (ERD) secondary to a vasoproliferative tumor (VPT) in an eye with sarcoidosis-associated intermediate uveitis. A 55-year-old woman previously diagnosed with sarcoidosis presented with decreased vision in the left eye (LE). Visual acuity in the LE was counting fingers. She had active vitritis, and a peripheral retinal vascular mass was noted in the superotemporal periphery. The mass was associated with ERD involving the posterior pole. The patient was managed with systemic and intravitreal steroids, and cyclosporine was subsequently added as a steroid-sparing agent. Because of recurrence of ERD, the patient underwent pars plana vitrectomy, and cryotherapy and laser photocoagulation were applied to the VPT. Two months postoperatively, visual acuity in the LE improved to 6/10. There was marked regression of the VPT and total resolution of the ERD. In conclusion, we report a favorable visual and clinical outcome in a patient with VPT-associated ERD who responded to a combination of medical therapy and surgical intervention. VPT may lead to different remote complications, so timely diagnosis of these tumors and proper management of their complications is warranted.

**Keywords:** Vasoproliferative tumor, sarcoidosis, intermediate uveitis, pars planitis

**Cite this article as:** Abdel Jalil S, Jaouni T, Amer R. Vasoproliferative Tumor Secondary to Sarcoidosis-Associated Intermediate Uveitis. Turk J Ophthalmol 2024;54:108-111

**Address for Correspondence:** Sara Abdel Jalil, Hadassah Medical Center, Department of Ophthalmology, Jerusalem, Israel
**E-mail:** dr.sara.jalil@gmail.com **ORCID-ID:** orcid.org/0009-0009-5150-3816

**Received:** 22.08.2023 **Accepted:** 31.01.2024

**DOI:** 10.4274/tjo.galenos.2024.36926

## Introduction

Vasoproliferative tumors (VPTs) are uncommon retinal lesions that may be primary or associated with other ocular conditions (secondary). Secondary VPTs most commonly occur in association with retinitis pigmentosa, pars planitis, and toxoplasmosis.[1]

VPTs can lead to different complications such as cystoid macular edema, exudative retinal detachment (ERD), and epiretinal membrane formation. Different modalities including surgery, cryotherapy, and radiotherapy have been used to treat these lesions.[1]

VPTs were also reported as one of the infrequent late complications in patients with intermediate uveitis (IU).[2]

Sarcoidosis is a systemic granulomatous inflammatory disease of unknown etiology. Its most common ocular manifestation is uveitis, reported in 25-50% of cases.[3]

Herein we report the clinical course and visual outcome of a middle-aged woman with inactive systemic sarcoidosis in whom chronic IU culminated in the development of VPT-induced ERD.

## Case Report

A 55-year-old woman was referred to the uveitis clinic because of localized ERD in her left eye (LE). She was diagnosed with pulmonary sarcoidosis by transbronchial biopsy four years earlier and treated with oral steroids for a period of one year. Subsequently she was in remission for three years prior to presentation. On examination, best corrected visual acuity (BCVA) was 6/15 in her right eye (RE) and counting fingers at 1 meter in the LE. She was pseudophakic in both eyes. RE anterior and posterior segments were normal. LE biomicroscopic exam revealed clear cornea and +2 anterior chamber flare. On LE fundoscopy, there was vitritis that obscured the posterior pole and a peripheral yellow vascular retinal mass was noted in the superotemporal periphery (Figure 1A). Fluorescein angiography

(FA) demonstrated signs of diffuse retinal vasculitis, macular and optic disc leakage, as well as leakage from the peripheral retinal mass (Figure 2). There were adjacent hard exudates and ERD that involved the macula (Figure 3A). B-scan ultrasound revealed the presence of vitreous opacities and a homogenous hyperechogenic retinal lesion with a thickness of 1.5 mm (Figure 4).

The patient was diagnosed as having sarcoidosis-associated chronic IU with ERD secondary to VPT. Treatment was initiated with pulse intravenous methylprednisolone (Upjohn [now Pfizer], Pennsylvania, USA) (1 g/day for 5 days) in addition to intravitreal triamcinolone acetonide (Bristol Myers Squibb, New Jersey, USA) (4 mg/0.1 mL). Subsequently, she received prednisone (Jubilant Cadista Pharmaceuticals, Salisbury, USA) and cyclosporine (Sandoz, Basel, Switzerland). The ERD totally resolved (Figure 3B). However, the patient did not perceive any improvement because of the persistent dense vitreous opacities and low LE BCVA (counting fingers at 3 meters). Seven months later, during prednisone tapering (at the dose of 5 mg/per day), the patient experienced recurrence of ERD involving the posterior pole. Intravitreal triamcinolone acetate (2 mg/0.05 mL) was administered. Ten days later, she received a preoperative pulse of intravenous methylprednisolone (1 g/day for 5 days). She then underwent pars plana vitrectomy because of the dense vitreous opacities, and cryotherapy and laser photocoagulation were applied to the VPT. Intravitreal triamcinolone acetate (2 mg/0.05 mL) was injected at the end of the operation. Two months postoperatively, LE BCVA improved to 6/10. There was marked regression of the VPT and of the adjacent hard exudates with complete resolution of the ERD (Figure 1B).

## Discussion

We herein report the clinical outcome of ERD secondary to VPT in a patient with sarcoidosis-associated chronic IU. The patient presented four years after the diagnosis of pulmonary sarcoidosis at a time when the systemic disease was quiescent. The patient had irregular follow-up prior to presentation and had not been properly treated for IU and ERD despite being previously diagnosed. Combined medical therapy and surgical intervention proved to be successful in the management of this case and yielded a favorable visual outcome. The presence of non-resolving vitreous opacities in the visual axis necessitated vitrectomy in addition to cryotherapy and laser photocoagulation.

The histopathological findings in IU-associated VPTs were first described by Henkind and Morgan[4] in 1964. They described them as lesions with a "Coats-like" phenotype in eyes that were enucleated due to malignant glaucoma. In 1983, Shields et al.[5] reported the presence of an unusual retinal vascular lesion in 12 eyes with ERD and called these tumors "presumed acquired non-familial retinal hemangioma". A decade later, Shields et al.[1] reported on 129 eyes with vascular lesions and suggested the term VPT. In their report, VPT was classified as secondary in 29 eyes, 8 of which were due to pars planitis. This vascular lesion

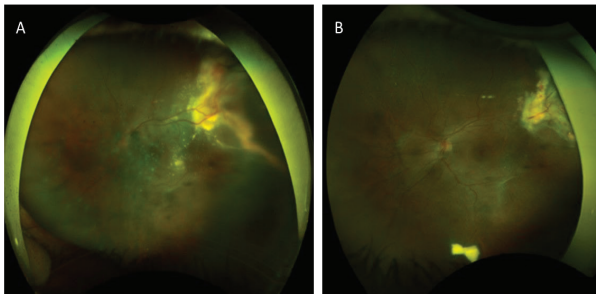


**Figure 1.** (A) Widefield pseudo-color fundus photograph (Optos Panoramic 200MA; Optos PLC, Dunfermline, Scotland, United Kingdom) of the left eye at presentation shows dense central vitritis obscuring the posterior pole and a peripheral temporal retinal vascular lesion with adjacent hard exudates. (B) Postoperatively, the vasoproliferative tumor is decreased in size, with marked resolution of the hard exudates. A clear view of the posterior pole is noted and the deposit of intravitreal triamcinolone acetonide is observed in inferior vitreous cavity

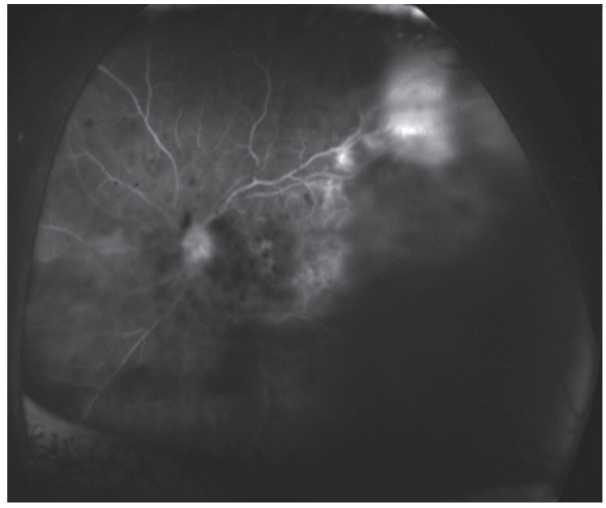

**Figure 2.** Fluorescein angiography of the left eye at presentation shows signs of vasculitis and leakage from the vasoproliferative tumor in the superotemporal periphery as well as from the optic disc and macula

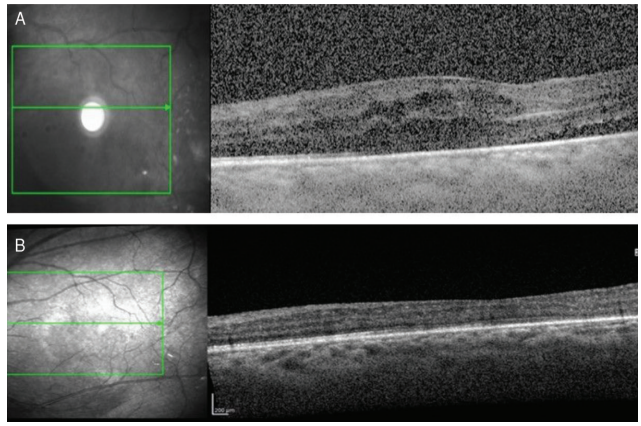


**Figure 3.** Upper panel A: optical coherence tomography (OCT) of the left eye at presentation shows subretinal and intraretinal fluid. Lower panel B: OCT of the same eye 3 weeks postoperatively shows resolution of the cystoid macular edema and exudative retinal detachment

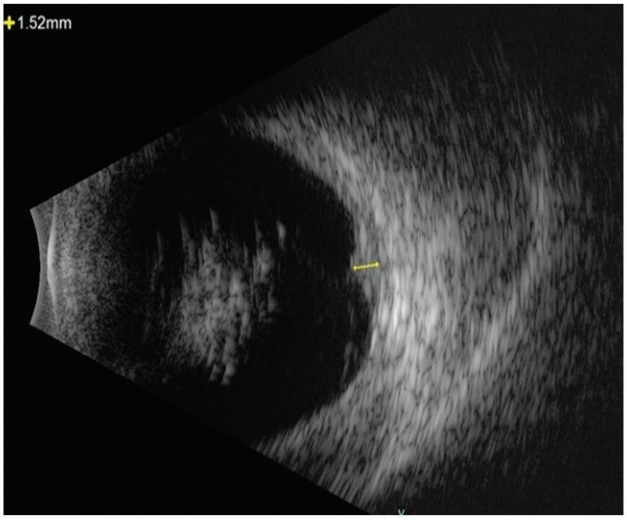


**Figure 4.** Ocular ultrasound (B-mode) of left eye at presentation revealed the presence of dense vitreous opacities and a homogenous hyperechogenic retinal lesion with a thickness of 1.5 mm (yellow mark)

was revealed to involve not only the retina, but infrequently the retinal pigment epithelium as well as choroid, so it was termed VPT of the ocular fundus.[1]

In 2013, Shields et al.[6] published a report of 275 patients with VPT (mean presenting age of 44 years). They reported that VPT predominantly affected females (59%) and was primary in 80% of patients and secondary in 20% of patients. The two most common causes of secondary VPT were retinitis pigmentosa (22%) and pars planitis (21%). Additional causes included Coats disease (16%) and previous retinal detachment surgery (12%).[6]

Although VPT is a benign lesion, it may lead to severe visual morbidity because of the distant effects of this tumor. According to the most recent report by Shields et al.[6], ocular complications include macular edema (32%), epiretinal membrane (20%) and vitreous hemorrhage (19%). Retinal exudation developed in 71% of cases (extramacular 48%, macular 23%).

IU is an inflammatory condition affecting predominantly young adults and children. It can affect either healthy individuals or it can occur secondary to other systemic diseases such as tuberculosis and multiple sclerosis. It is usually bilateral.[7] This type of uveitis is characterized by vitritis and a relatively quiet anterior chamber. IU may be associated with diverse ocular complications of the anterior and/or posterior segment.[2] Because of the wide spectrum of associated vision-threatening sequelae, it may lead to a potentially guarded prognosis.[7] A cohort study including 96 patients (174 eyes) with IU reported on its early and long-term complications.[2] VPT was among the late complications, described in 1% of eyes. The mean time between uveitis diagnosis and the development of secondary VPT was found to be 160 months.[6]

The pathogenesis of VPT in eyes with IU remains unknown. It was proposed that inadequately controlled inşammation, peripheral vascular leakage, and hypoxic alterations in the pars plana region may lead to retinal elevations and VPT. In a multicentric study, Pollack et al.[8] reported 13 eyes with pars planitis who also had peripheral retinal elevation. All of these eyes showed signs of chronic inflammation, including the presence of snow-banking, inferior fibrovascular abnormalities, and/or marked RPE hypertrophy.

A recent multicentric study of VPTs secondary to IU reported on 36 eyes of 34 patients (22 females).[9] The mean age at onset was 35 years. Twenty-nine patients were diagnosed with pars planitis, three patients had tuberculosis, and one had multiple sclerosis. The VPTs were unilateral in 93.7% of the cases. At the time of VPT diagnosis, all these eyes had active IU, which was defined by the presence of vitritis and/or vascular leakage on FA.[9]

Retinal VPTs secondary to IU are relatively rare tumors, which is the reason for the lack of an evidence-based consensus regarding the best way to treat these lesions. Pichi et al.[9] reported the results of local treatment in 22 of 36 eyes with VPT secondary to IU. Treatment consisted of cryotherapy (8 eyes), argon laser photocoagulation (10 eyes), intravitreal anti-VEGF (2 eyes), or a combination of anti-VEGF injections and cryotherapy (2 eyes). The other 14 eyes were observed without receiving any direct treatment for these tumors. During the follow-up period, the treated VPTs had a decrease in tumor thickness to 1.25 mm (mean initial thickness was 3.13 mm). In the untreated group, the final mean tumor thickness remained stable at 2.65 mm.

In conclusion, VPTs need to be managed with a variety of modalities, particularly in the setting of uveitis. Awareness and recognition of these tumors with proper management of their complications is needed.

### Ethics

**Informed Consent:** Obtained.

### Authorship Contributions

Surgical and Medical Practices: T.J., R.A., Concept: R.A., Design: S.A.J., R.A., Data Collection or Processing: S.A.J., Analysis or Interpretation: R.A., Literature Search: S.A.J., Writing: S.A.J., R.A.

**Conflict of Interest:** No conflict of interest was declared by the authors.

**Financial Disclosure:** The authors declared that this study received no financial support.

## References

1. Shields CL, Shields JA, Barrett J, De Potter P. Vasoproliferative tumors of the ocular fundus. Classification and clinical manifestations in 103 patients. Arch Ophthalmol. 1995;113:615-623.
2. Sancho L, Kramer M, Koriat A, Eiger-Moscovich M, Sharon Y, Amer R. Complications in Intermediate Uveitis: Prevalence, Time of Onset, and Effects on Vision in Short-Term and Long-Term Follow-Up. Ocul Immunol Inflamm. 2019;27:447-455.
3. Jamilloux Y, Kodjikian L, Broussolle C, Sève P. Sarcoidosis and uveitis. Autoimmun Rev. 2014;13:840-849.
4. Henkind P, Morgan G. Peripheral retinal angioma with exudative retinopathy in adults (coats's lesion). Br J Ophthalmol. 1966;50:2-11.
5. Shields JA, Decker WL, Sanborn GE, Augsburger JJ, Goldberg RE. Presumed acquired retinal hemangiomas. Ophthalmology. 1983;90:1292-1300.

6. Shields CL, Kaliki S, Al-Dahmash S, Rojanaporn D, Shukla SY, Reilly B, Shields JA. Retinal vasoproliferative tumors: comparative clinical features of primary vs secondary tumors in 334 cases. JAMA Ophthalmol. 2013;131:328-334.
7. Ozdal PC, Berker N, Tugal-Tutkun I. Pars Planitis: Epidemiology, Clinical Characteristics, Management and Visual Prognosis. J Ophthalmic Vis Res. 2015;10:469-480.
8. Pollack AL, McDonald HR, Johnson RN, Ai E, Irvine AR, Lahey JM, Lewis H, Rodriguez A, Ryan EH Jr, Shields CL. Peripheral retinoschisis and exudative retinal detachment in pars planitis. Retina. 2002;22:719-724.
9. Pichi F, Neri P, Agarwal A, Invernizzi A, Choudhry N, Amer R, Lembo A, Nucci P, Thompson I, Sen HN, Shields CL. Vasoproliferative Tumors In Intermediate Uveitis. Retina. 2020;40:1765-1773.