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Retinal vasoproliferative tumor (VPT) is a histopathologically benign vascularized glial tumor believed to result from the proliferation of blood vessels, glial tissue, and retinal pigment epithelium (RPE). VPT was first described in 1983 under the term presumed acquired retinal hemangioma.\(^1\)\(^-\)\(^3\) This tumor generally appears in the fourth and fifth decade of life with features of a peripheral yellow-red intraretinal mass most often located temporally or inferotemporally.\(^4\) VPTs are classified into primary (idiopathic, 80%) or secondary (20%) types.\(^4\)

Primary VPTs are deemed idiopathic in otherwise healthy eyes, whereas secondary VPTs have been associated with preexisting congenital, inflammatory, vascular, traumatic, and dystrophic retinal conditions.\(^4\) Secondary forms are more often bilateral, multiple, and accompanied by poorer visual acuity.\(^4\) Both types of VPTs are benign, but either can compromise vision due to retinal exudation, retinal detachment, intraretinal edema, membrane formation, and vitreous hemorrhage.\(^5\) Although these tumors are benign, it is important to identify and treat them to avoid visual compromise. In this report, we describe a patient with bilateral retinitis pigmentosa (RP) who developed secondary VPT.

CASE REPORT

A 39-year-old white woman who had been diagnosed with bilateral RP at age 26 was referred for evaluation of an intraocular mass. The lesion was initially considered to be a choroidal neovascular membrane (CNVM) and had been treated elsewhere with laser photocoagulation 8 months previously.

At the time of presentation at our center, the patient’s visual acuity was 20/25 in the right eye (OD) and 20/30 in the left eye (OS). The anterior segment was unremarkable in each eye. Her fundus examination showed bilateral attenuated retinal vasculature and 360° bone-spicule pigmentation within the retina, compatible with the previous diagnosis of RP. Additionally, a peripheral elevated orange-red retinal mass was noted OS, surrounded by a shallow, dependent exudative retinal detachment and measuring 4.5 x 4.0 x 2.5 mm (Figure 1A). Fluorescein angiography (FA) demonstrated rapid filling of the tumor from a nondilated feeding retinal artery and late leakage (Figure 1B). B-scan ultrasonography showed a dense mass (Figure 2A), and spectral-domain optical coherence tomography (SD-OCT) showed normal foveal contour OD and a blunted foveal depression OS secondary to an epiretinal membrane (ERM) (Figure 2B).

**AT A GLANCE**

- Retinal vasoproliferative tumors (VPTs) typically develop in individuals in their 30s and 40s.
- Primary VPTs are deemed idiopathic in otherwise healthy eyes, whereas secondary VPTs have been associated with preexisting congenital, inflammatory, vascular, traumatic, and dystrophic retinal conditions. Secondary forms are more often bilateral, multiple, and accompanied by poorer visual acuity.
- Both types of VPTs are benign, but either can compromise vision due to retinal exudation, retinal detachment, intraretinal edema, membrane formation, and vitreous hemorrhage.
- Management of VPTs depends on tumor features and patient symptoms.
SD-OCT over the mass confirmed a full-thickness intraretinal mass with superficial intralesional cysts (Figure 2C). The clinical picture was compatible with an active secondary VPT in the setting of RP.

Management options were discussed with the patient, including surrounding laser photocoagulation, indocyanine green–enhanced transpupillary thermotherapy (ICG-enhanced TTT), verteporfin (Visudyne, Bausch + Lomb) photodynamic therapy (PDT), and cryotherapy. PDT was performed with standard settings and with simultaneous intravitreal anti-VEGF therapy. Four months later, the tumor had responded with slow involution, partial resolution of surrounding subretinal fluid and exudation, and thinning of the retinal feeder vessel.

DISCUSSION

RP, which affects approximately 1 in 4,000 persons worldwide, refers to a group of inherited disorders in which degenerative abnormalities of the photoreceptors lead to progressive vision loss. Exudative vasculopathy manifesting as Coats-like disease, VPT, or CNVM complicates approximately 5% of RP cases. The pathophysiology of the different manifestations of related exudative vasculopathy might result from chronic retinal ischemia from attenuation of the retinal vessels or exudation from incompetent retinal vessels. Additionally, chronic damage in the RPE could provide access for secondary choroidal-retinal anastomoses through Bruch membrane defects.

In a large series of 334 cases of VPT in 275 patients, 80% (n = 219) of patients had primary and 20% (n = 56) had secondary VPT (see infographic at right). In the 56 patients with secondary tumors, underlying ocular diseases included RP (n = 15, 22%), pars planitis (n = 14, 21%), Coats disease (n = 11, 16%), previous retinal detachment surgery (n = 8, 12%), and idiopathic peripheral retinal vasculitis (n = 4, 6%). A comparison of primary versus secondary VPT revealed significant differences, in that primary tumors occurred more often at older age (46 vs. 38 years) and with less frequency of visual symptoms (74% vs. 87%), visual acuity <20/200 (15% vs. 28%), bilaterality (4% vs. 20%), and multifocality (5% vs. 15%).

![Figure 1](image1.png)

Figure 1. A 39-year-old woman with RP and retinal VPT misdiagnosed as neovascular membrane and treated at another center with laser photocoagulation. Fundus OS shows typical RP changes and a yellow-red mass located inferotemporally, posterior to the equator and surrounded by dense exudation. Note the minimally dilated feeding retinal vessels (A). Late-phase FA demonstrates diffuse leakage from the tumor (B).

![Figure 2](image2.png)

Figure 2. Imaging of VPT in a patient with RP. B-scan ultrasonography discloses a dome-shaped retinal mass measuring 4.5 x 4.0 x 2.5 mm (A). SD-OCT displays a discrete blunted foveal contour secondary to an ERM (B). SD-OCT over the lesion demonstrates a full thickness retinal mass with two intralesional cystic spaces that could represent intralesional fluid or large vessels (C).
In addition to tumor control, cryotherapy can lead to release of ERM in 63% of [VPT] cases.