Retinal Vasoproliferative Tumors

Comparative Clinical Features of Primary vs Secondary Tumors in 334 Cases

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Objective: To compare the clinical features of primary vs secondary retinal vasoproliferative tumors (VPTs).

Methods: Retrospective case series of 334 tumors in 295 eyes of 275 patients.

Results: Of 275 patients with VPT, 41% (n=113) were male and 59% (n=162) were female, with a mean age of 44 years at presentation. Primary VPT occurred in 80% (n=219) and secondary VPT, in 20% (n=56) of patients. Secondary VPT (n=67) occurred in eyes with retinitis pigmentosa (n=15, 22%), pars planitis (n=14, 21%), Coats disease (n=11, 16%), previous retinal detachment surgery (n=8, 12%), idiopathic peripheral retinal vasculitis (n=4, 6%), familial exudative vitreoretinopathy (n=3, 4%), and others (n=12, 18%). The mean interval between diagnosis of underlying ocular condition and secondary VPT was 160 months. Statistically significant differences (P<.05) in clinical features (primary vs secondary VPTs) included mean age at presentation (46 vs 38 years), visual symptoms (74% vs 87%), poor visual acuity worse than 20/200 (15% vs 28%), bilaterality (4% vs 20%), multifocality (5% vs 15%), postequatorial tumor location (20% vs 33%), tumor basal dimension (6 vs 7 mm), anterior chamber cells (16% vs 30%), and vitreous cells (19% vs 48%).

Conclusions: Retinal vasoproliferative tumor can be primary (80%) or secondary (20%). Compared with primary VPT, secondary VPT is more often bilateral, multiple, and larger and occurs at an earlier age associated with poorer visual acuity.


METHODS

The medical records of all patients with the clinical diagnosis of VPT of the ocular fundus or acquired retinal hemangioma diagnosed between March 26, 1975, and November 30, 2011, were retrospectively reviewed. Institutional review board approval was obtained. Data were retrospectively collected from medical record review regarding patient demographics includ-
ing age (in years), race (white, African American, Hispanic, or Asian), sex (male or female), medical history (hypertension, diabetes mellitus, hypercholesterolemia, autoimmune disease, oculoneurocutaneous syndromes [phakomatoses], systemic steroid use, and cigarette use), and ocular history (anterior segment/posterior segment disease, topical/intravitreal medication, and ocular surgery).

The clinical data included presenting symptoms, duration of symptoms, visual acuity, intraocular pressure (in millimeters of mercury), laterality (unilateral or bilateral), and anterior segment findings. The tumor features included number of tumors, tumor size (basal dimension and thickness in millimeters), and tumor location by quadrant, anteroposterior region, proximity to the optic disc (in millimeters), and proximity to the foveola (in millimeters). Other tumor features included color and vascularity. Associated vitreoretinal findings of vitreous cells, vitreous hemorrhage, retinal pigment epithelial alterations, subretinal/intraretinal exudation and hemorrhage, subretinal fluid, epiretinal membrane, cystoid macular edema, and retinal/optic disc neovascularization were recorded.

The anterior segment findings were confirmed by slitlamp examination and documented by large anterior segment drawings and slitlamp photography. The posterior segment findings were confirmed on ophthalmoscopic examination and documented by large fundus drawings, fundus photography, fluorescein angiography, ultrasonography, and optical coherence tomography.

The clinical features were evaluated based on primary vs secondary VPTs. Primary VPT was defined as VPT with no evident previous ocular condition, whereas secondary VPT was defined as VPT occurring in an eye with a known or clinically evident underlying ocular condition. All proportions in each group were presented as number and percentage and comparison between the groups was provided using the Fisher exact test or χ² test. The data collected on continuous or ordinal scale were expressed as mean, median, minimum, and maximum. An independent-samples t test was performed to compare age, intraocular pressure, tumor base, tumor thickness, and proximity to the optic nerve and foveola as a function of primary vs secondary VPTs.

### RESULTS

There were 334 VPTs in 295 eyes of 275 patients. The demographic data and clinical features are listed in Table 1. The table displays the number and percentage of patients with primary and secondary VPTs, as well as the comparison of visual acuity, intraocular pressure, tumor size, and other clinical features between the two groups.

**Table 1. Demographics and Ocular Features of 295 Eyes of 275 Patients With 334 VPTs**

<table>
<thead>
<tr>
<th>Feature</th>
<th>Primary VPT (n = 252 Tumors in 228 Eyes of 219 Patients)</th>
<th>Secondary VPT (n = 82 Tumors in 67 Eyes of 56 Patients)</th>
<th>All VPT (n = 334 Tumors in 295 Eyes of 275 Patients)</th>
<th>P Value in Comparison of Primary vs Secondary VPTa</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age, y (n = 271 patients)</td>
<td>Mean 46</td>
<td>38</td>
<td>44</td>
<td>.005b</td>
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<tr>
<td></td>
<td>Median (range) 47 (2-87)</td>
<td>37 (1-82)</td>
<td>45 (1-87)</td>
<td></td>
</tr>
<tr>
<td>Sex (n = 275 patients)</td>
<td>M 86 (39)</td>
<td>27 (48)</td>
<td>113 (41)</td>
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</tr>
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<td></td>
<td>F 133 (61)</td>
<td>29 (52)</td>
<td>162 (59)</td>
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</tr>
<tr>
<td>Race (n = 275 patients)</td>
<td>White 180 (82)</td>
<td>45 (80)</td>
<td>225 (82)</td>
<td>.75c</td>
</tr>
<tr>
<td></td>
<td>African American 18 (8)</td>
<td>6 (11)</td>
<td>24 (9)</td>
<td></td>
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<tr>
<td></td>
<td>Hispanic 12 (5)</td>
<td>4 (7)</td>
<td>16 (6)</td>
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<tr>
<td></td>
<td>Asian 9 (4)</td>
<td>1 (2)</td>
<td>10 (4)</td>
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<tr>
<td>Systemic disease (n = 261 patients)</td>
<td>Hypertension 44 (20)</td>
<td>9 (17)</td>
<td>53 (20)</td>
<td>.57</td>
</tr>
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<td></td>
<td>Diabetes mellitus 11 (5)</td>
<td>1 (2)</td>
<td>12 (5)</td>
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<td></td>
<td>Hypercholesterolemia 27 (13)</td>
<td>6 (11)</td>
<td>33 (13)</td>
<td>.82</td>
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<td>Neurofibromatosis type 1 2 (1)</td>
<td>0 (0)</td>
<td>2 (&lt;1)</td>
<td>&gt;.99</td>
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<td>Autoimmune disease 6 (3)</td>
<td>3 (6)</td>
<td>9 (3)</td>
<td>.40</td>
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<tr>
<td>Symptoms (n = 314 tumors)</td>
<td>Asymptomatic 62 (26)</td>
<td>10 (13)</td>
<td>72 (23)</td>
<td>&lt;.001c</td>
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<tr>
<td></td>
<td>Decreased vision/visual field defect 124 (52)</td>
<td>48 (63)</td>
<td>172 (55)</td>
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<td>Floaters/floats of light 41 (17)</td>
<td>10 (13)</td>
<td>51 (16)</td>
<td>.02</td>
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<td></td>
<td>Decreased vision and floaters 9 (4)</td>
<td>1 (1)</td>
<td>10 (3)</td>
<td></td>
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<td></td>
<td>Others 2 (&lt;1)</td>
<td>7 (9)</td>
<td>9 (3)</td>
<td></td>
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<tr>
<td>Laterality (n = 275 patients)</td>
<td>Unilateral 210 (96)</td>
<td>45 (80)</td>
<td>255 (93)</td>
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<tr>
<td></td>
<td>Bilateral 9 (4)</td>
<td>11 (20)</td>
<td>20 (7)</td>
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<tr>
<td>Visual acuity (n = 284 eyes)</td>
<td>20/20 to 20/50</td>
<td>30 (44)</td>
<td>173 (61)</td>
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<tr>
<td></td>
<td>20/60 to 20/200</td>
<td>19 (28)</td>
<td>60 (21)</td>
<td>.004c</td>
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<tr>
<td></td>
<td>&lt;20/200</td>
<td>19 (28)</td>
<td>51 (18)</td>
<td></td>
</tr>
<tr>
<td>Intraocular pressure, mm Hg (n = 284 eyes)</td>
<td>Mean 16</td>
<td>16</td>
<td>16</td>
<td>.25b</td>
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<tr>
<td></td>
<td>Median (range) 16 (6-45)</td>
<td>14 (10-45)</td>
<td>15 (6-45)</td>
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</tr>
</tbody>
</table>

Abbreviation: VPT, vasoproliferative retinal tumor.

a Fisher exact test.

b Independent-samples t test.

χ² Test.

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The mean age at presentation was 44 years (median, 45 years; range, 1-87 years). A comparison (primary vs secondary VPTs) revealed statistically significant ($P < .05$) clinical features including mean age at presentation (46 vs 38 years), visual symptoms (74% vs 87%), bilaterality (4% vs 20%), and poor visual acuity worse than 20/200 (15% vs 28%) (Table 1).

Table 2. Clinical Tumor and Related Features of 295 Eyes of 275 Patients With 334 VPTs

| Feature                                      | Primary VPT (n = 252 Tumors in 22 Eyes of 219 Patients) | Secondary VPT (n = 82 Tumors in 67 Eyes of 56 Patients) | All VPT (n = 334 Tumors in 295 Eyes of 275 Patients) | $P$ Value in Comparison of Primary vs Secondary VPT
<table>
<thead>
<tr>
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<tr>
<td>Anterior segment findings</td>
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<tr>
<td>Cataract (n = 206 eyes)</td>
<td>51 (22)</td>
<td>25 (33)</td>
<td>76 (25)</td>
<td>.09</td>
</tr>
<tr>
<td>Glaucoma (n = 291 eyes)</td>
<td>10 (4)</td>
<td>2 (3)</td>
<td>12 (4)</td>
<td>.74</td>
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<tr>
<td>Anterior chamber cell/flare (n = 273 eyes)</td>
<td>44 (19)</td>
<td>24 (32)</td>
<td>68 (22)</td>
<td>.03</td>
</tr>
<tr>
<td>Iris neovascularization (n = 292 eyes)</td>
<td>9 (4)</td>
<td>2 (3)</td>
<td>11 (4)</td>
<td>&gt;.99</td>
</tr>
<tr>
<td>No. of VPTs per eye (n = 294 eyes)</td>
<td></td>
<td></td>
<td></td>
<td></td>
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<tr>
<td>Mean</td>
<td>1.1</td>
<td>1.2</td>
<td>1</td>
<td>.09</td>
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<tr>
<td>Median (range)</td>
<td>1 (1-5)</td>
<td>1 (1-3)</td>
<td>1 (1-5)</td>
<td>.09</td>
</tr>
<tr>
<td>No. of VPTs per eye (n = 294 eyes)</td>
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<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Single</td>
<td>217 (95)</td>
<td>56 (85)</td>
<td>273 (92)</td>
<td>.01</td>
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<tr>
<td>Multiple</td>
<td>11 (5)</td>
<td>10 (15)</td>
<td>21 (7)</td>
<td>.01</td>
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<td>Quadrant tumor location (n = 325 tumors)</td>
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</tr>
<tr>
<td>Macula</td>
<td>5 (2)</td>
<td>3 (4)</td>
<td>8 (2)</td>
<td>.03</td>
</tr>
<tr>
<td>Inferotemporal</td>
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<tr>
<td>Macula to equator</td>
<td>51 (20)</td>
<td>27 (33)</td>
<td>78 (24)</td>
<td>.02</td>
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<td>Equator to ora serrata</td>
<td>191 (77)</td>
<td>51 (63)</td>
<td>242 (73)</td>
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<tr>
<td>Anterior to ora serrata</td>
<td>2 (&lt;1)</td>
<td>0</td>
<td>2 (&lt;1)</td>
<td>.01</td>
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<td>Largest tumor basal diameter, mm (n = 297 tumors)</td>
<td>6 (0.5-15)</td>
<td>6 (0.5-25)</td>
<td>6 (0.5-25)</td>
<td>&lt;.001</td>
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<td>Tumor thickness, mm (n = 261 tumors)</td>
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</tr>
<tr>
<td>Mean</td>
<td>2.8</td>
<td>3</td>
<td>2.9</td>
<td>.26</td>
</tr>
<tr>
<td>Median (range)</td>
<td>2.6 (0-9.7)</td>
<td>2.9 (0-7.5)</td>
<td>2.6 (0-9.7)</td>
<td>.35</td>
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<td>Distance to optic nerve, mm (n = 310 tumors)</td>
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</tr>
<tr>
<td>Mean</td>
<td>12</td>
<td>11</td>
<td>12</td>
<td>.07</td>
</tr>
<tr>
<td>Median (range)</td>
<td>12 (0-20)</td>
<td>13 (0-20)</td>
<td>12 (0-20)</td>
<td>.07</td>
</tr>
<tr>
<td>Distance to foveola, mm (n = 310 tumors)</td>
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<td></td>
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<tr>
<td>Mean</td>
<td>11</td>
<td>10</td>
<td>11</td>
<td>.07</td>
</tr>
<tr>
<td>Median (range)</td>
<td>12 (0-20)</td>
<td>11 (0-17)</td>
<td>12 (0-20)</td>
<td>.07</td>
</tr>
<tr>
<td>Associated vitreoretinal findings</td>
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<tr>
<td>Vitreous cells (n = 329)</td>
<td>48 (19)</td>
<td>39 (48)</td>
<td>87 (26)</td>
<td>&lt;.001</td>
</tr>
<tr>
<td>Vitreous hemorrhage (n = 328)</td>
<td>44 (18)</td>
<td>18 (22)</td>
<td>62 (19)</td>
<td>.42</td>
</tr>
<tr>
<td>Epiretinal membrane (n = 330 tumors)</td>
<td>50 (20)</td>
<td>15 (18)</td>
<td>65 (20)</td>
<td>.75</td>
</tr>
<tr>
<td>Macula involved</td>
<td>33 (13)</td>
<td>6 (7)</td>
<td>39 (12)</td>
<td>.17</td>
</tr>
<tr>
<td>Macula not involved</td>
<td>17 (7)</td>
<td>9 (11)</td>
<td>26 (8)</td>
<td>.07</td>
</tr>
<tr>
<td>Dilated feeder vessels (n = 323 tumors)</td>
<td>139 (57)</td>
<td>45 (56)</td>
<td>184 (57)</td>
<td>.80</td>
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<td>Retinoschisis (n = 3)</td>
<td>4 (3)</td>
<td>3 (5)</td>
<td>7 (4)</td>
<td>.43</td>
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<tr>
<td>Subretinal fluid (n = 328 tumors)</td>
<td>142 (58)</td>
<td>42 (51)</td>
<td>184 (56)</td>
<td>.31</td>
</tr>
<tr>
<td>Macula involved</td>
<td>60 (24)</td>
<td>18 (22)</td>
<td>78 (24)</td>
<td>.77</td>
</tr>
<tr>
<td>Macula not involved</td>
<td>82 (33)</td>
<td>24 (29)</td>
<td>106 (32)</td>
<td>.65</td>
</tr>
<tr>
<td>Tractional retinal detachment (n = 327 tumors)</td>
<td>10 (4)</td>
<td>7 (9)</td>
<td>17 (5)</td>
<td>.15</td>
</tr>
<tr>
<td>Macula involved</td>
<td>3 (1)</td>
<td>4 (5)</td>
<td>7 (2)</td>
<td>.07</td>
</tr>
<tr>
<td>Macula not involved</td>
<td>7 (3)</td>
<td>3 (4)</td>
<td>10 (3)</td>
<td>.07</td>
</tr>
<tr>
<td>Subretinal/intraretinal exudation (n = 331 tumors)</td>
<td>179 (72)</td>
<td>56 (68)</td>
<td>235 (71)</td>
<td>.58</td>
</tr>
<tr>
<td>Macula involved</td>
<td>55 (22)</td>
<td>20 (24)</td>
<td>75 (23)</td>
<td>.65</td>
</tr>
<tr>
<td>Macula not involved</td>
<td>124 (50)</td>
<td>36 (44)</td>
<td>160 (48)</td>
<td>.35</td>
</tr>
<tr>
<td>Subretinal/intraretinal hemorrhage (n = 330 tumors)</td>
<td>94 (38)</td>
<td>20 (24)</td>
<td>114 (35)</td>
<td>.03</td>
</tr>
<tr>
<td>Retinal pigment epithelial alterations (n = 323 tumors)</td>
<td>65 (27)</td>
<td>40 (50)</td>
<td>105 (33)</td>
<td>&lt;.001</td>
</tr>
<tr>
<td>Retinal neovascularization (n = 327 eyes)</td>
<td>4 (2)</td>
<td>1 (1)</td>
<td>5 (2)</td>
<td>&gt;.99</td>
</tr>
</tbody>
</table>

Abbreviation: VPT, vasoproliferative retinal tumor.

a In some cases, the features were not visible because of media opacity or were not recorded in the medical record.

b Fisher exact test.

c $\chi^2$ Test.

d Independent-samples t test.

Table 1. The mean age at presentation was 44 years (median, 45 years; range, 1-87 years). A comparison (primary vs secondary VPTs) revealed statistically significant ($P < .05$) clinical features including mean age at presentation (46 vs 38 years), visual symptoms (74% vs 87%), bilaterality (4% vs 20%), and poor visual acuity worse than 20/200 (15% vs 28%) (Table 1).
mor basal dimension (6 vs 7 mm), anterior chamber cells (16% vs 30%), and vitreous cells (19% vs 48%) (Table 2). Similar features (primary vs secondary VPTs) included tumor location in the inferotemporal quadrant (66% vs 72%), tumor location between the equator and ora serrata (77% vs 63%), and tumor thickness (2.8 vs 3 mm).

The results of imaging techniques are listed in Table 3. The tumor appeared ultrasonographically dense in 87% and hyperfluorescent in arterial (92%), venous (96%), and late (56%) phases of fluorescein angiography. Optical coherence tomography (n = 134 eyes) documented cystoid macular edema (32%). Ultrasonography (n = 248 eyes), fluorescein angiography (n = 219 eyes), and optical coherence tomography (n = 134 eyes) showed no statistical difference when comparing primary vs secondary tumors.

The associated ocular diseases in 67 eyes with secondary VPT are listed in Table 4. The 3 most common diseases included retinitis pigmentosa (22%), pars planitis (21%), and Coats disease (16%). Both eyes were involved in 11 cases (20%), and in this cohort, the underlying conditions included retinitis pigmentosa (n = 5), pars planitis (n = 3), aniridia (n = 1), idiopathic choroiditis (n = 1), and idiopathic peripheral retinal vasculitis (n = 1). The mean interval between diagnosis of the associated ocular disease and diagnosis of secondary VPT was 160 months (median, 93 months; range, 0-736 months).

**Table 3. Imaging Features of 295 Eyes of 275 Patients With 334 VPTs**

<table>
<thead>
<tr>
<th>Feature</th>
<th>Primary VPT (n = 252 Tumors in 228 Eyes of 219 Patients)</th>
<th>Secondary VPT (n = 82 Tumors in 67 Eyes of 56 Patients)</th>
<th>All VPT (n = 334 Tumors in 295 Eyes of 275 Patients)</th>
<th>P Value in Comparison of Primary vs Secondary VPTa</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Ultrasonography (n = 248 Eyes)</strong></td>
<td></td>
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<tr>
<td>Tumor configurationb</td>
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<td></td>
</tr>
<tr>
<td>Irregular</td>
<td>28 (15)</td>
<td>12 (22)</td>
<td>40 (16)</td>
<td>.27</td>
</tr>
<tr>
<td>Dome</td>
<td>158 (82)</td>
<td>43 (78)</td>
<td>201 (81)</td>
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<tr>
<td>Diffuse</td>
<td>4 (2)</td>
<td>0</td>
<td>4 (2)</td>
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<td><strong>Acoustic quality</strong></td>
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<tr>
<td>Dense</td>
<td>161 (85)</td>
<td>54 (93)</td>
<td>215 (87)</td>
<td>.12c</td>
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<tr>
<td>Hollow</td>
<td>29 (15)</td>
<td>4 (7)</td>
<td>33 (13)</td>
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<td><strong>Fluorescein Angiography (n = 219 Tumors)</strong></td>
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<tr>
<td>Arterial phase</td>
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<tr>
<td>Hyperfluorescence</td>
<td>146 (91)</td>
<td>48 (94)</td>
<td>194 (92)</td>
<td>.07</td>
</tr>
<tr>
<td>Hypofluorescence</td>
<td>13 (8)</td>
<td>1 (2)</td>
<td>14 (7)</td>
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<td>Normal fluorescence</td>
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<td>2 (4)</td>
<td>3 (1)</td>
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<td>Venous phase</td>
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<tr>
<td>Hyperfluorescence</td>
<td>155 (96)</td>
<td>51 (94)</td>
<td>206 (96)</td>
<td>.22</td>
</tr>
<tr>
<td>Hypofluorescence</td>
<td>5 (3)</td>
<td>1 (2)</td>
<td>6 (3)</td>
<td></td>
</tr>
<tr>
<td>Normal fluorescence</td>
<td>1 (&lt;1)</td>
<td>2 (4)</td>
<td>3 (1)</td>
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<td>Late phase</td>
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<tr>
<td>Hyperfluorescence</td>
<td>91 (57)</td>
<td>30 (56)</td>
<td>121 (56)</td>
<td>.36</td>
</tr>
<tr>
<td>Hypofluorescence</td>
<td>5 (3)</td>
<td>1 (2)</td>
<td>6 (3)</td>
<td></td>
</tr>
<tr>
<td>Normal fluorescence</td>
<td>0</td>
<td>1 (2)</td>
<td>1 (&lt;1)</td>
<td></td>
</tr>
<tr>
<td>Leakage</td>
<td>65 (40)</td>
<td>22 (41)</td>
<td>87 (40)</td>
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<tr>
<td><strong>Optical Coherence Tomography (n = 134 Eyes)</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Central foveal thickness</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Mean</td>
<td>341</td>
<td>354</td>
<td>345</td>
<td>.61d</td>
</tr>
<tr>
<td>Median (range)</td>
<td>317 (111-961)</td>
<td>350 (152-651)</td>
<td>323 (111-961)</td>
<td></td>
</tr>
<tr>
<td>Cystoid macular edema</td>
<td>28 (29)</td>
<td>15 (41)</td>
<td>43 (32)</td>
<td>.22c</td>
</tr>
</tbody>
</table>

Abbreviation: VPT, vasoproliferative retinal tumor.

a x² Test.
b Ultrasonography was performed in 248 eyes but tumor configuration was not mentioned or clearly shown in every case so that the numbers do not add up to the total number.
c Fisher exact test.
d Independent-samples t test.

Clinically, retinal VPT appears as a yellow-red, often ill-defined retinal-based mass with a mildly dilated feeding retinal artery and draining vein. Because of the classic peripheral location, indistinct margins, and color similar to the background fundus, the VPT can be somewhat difficult to visualize ophthalmoscopically (Figures 1, 2, and 3). Although VPT is a benign tumor, it can produce profound visual loss related to remote effects of the tumor, including macular exudation, cystoid macular edema, and vitreous hemorrhage and epimacular membrane. These complications led to visual acuity of 20/200 or worse in 18% of our cases. In this analysis, retinal exudation was found in 71% of cases, with involvement of the macula (23%) and extramacular (48%) regions. Macular edema was found in 32% and epiretinal membrane, in 20%.

**Table 4. Associated Ocular Diseases in 67 Eyes With Secondary VPTs**
Retinal VPT has been classified into primary (idiopathic) and secondary types. Primary VPTs are typically located in the inferotemporal (42%) or inferior (21%) portion of the fundus and tend to be solitary, small, and between the globe equator and ora serrata. Secondary VPTs are more often multifocal, bilateral, and believed to be a reactive vascular response to a variety of ocular insults. In our series of 67 eyes with secondary VPT, the underlying ocular conditions included retinitis pigmentosa (22%), pars planitis (21%), Coats disease (16%), previous retinal detachment repair (12%), and others (Table 4).

In this report, we have analyzed our experience with VPT to explore distinguishing characteristics of primary vs secondary tumors (Figure 1 and Figure 2). We found that primary VPT (compared with secondary VPT) showed statistically significant differences, with older age at presentation and less frequency of symptoms, bilaterality, poor visual acuity, and tumor multifocality. Primary VPT was less likely postequatorial in location and had smaller tumor basal dimension (Table 1 and Table 2). Despite these differences, there were many similarities of primary and secondary

### Table 4. Associated Ocular Conditions in 67 Eyes of 56 Patients With Secondary VPT

<table>
<thead>
<tr>
<th>Associated Ocular Condition</th>
<th>Secondary VPT, No. (%)</th>
<th>Interval From Diagnosis of Ocular Condition to Development of VPT, mo, Mean; Median (Range)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Eyes (n = 67)</td>
<td>Patients (n = 56)</td>
<td></td>
</tr>
<tr>
<td>Retinitis pigmentosa</td>
<td>15 (22)</td>
<td>228; 198 (0-564)</td>
</tr>
<tr>
<td>Pars planitis</td>
<td>14 (21)</td>
<td>112; 39 (0-540)</td>
</tr>
<tr>
<td>Coats disease</td>
<td>11 (16)</td>
<td>67; 3 (0-264)</td>
</tr>
<tr>
<td>Previous retinal detachment repair</td>
<td>8 (12)</td>
<td>477; 480 (216-736)</td>
</tr>
<tr>
<td>Idiopathic peripheral retinal vasculitis</td>
<td>4 (6)</td>
<td>0</td>
</tr>
<tr>
<td>Familial exudative vitreoretinopathy</td>
<td>3 (4)</td>
<td>316; 228 (0-720)</td>
</tr>
<tr>
<td>Toxplasmosis</td>
<td>3 (4)</td>
<td>300; 360 (96-444)</td>
</tr>
<tr>
<td>Aniridia</td>
<td>2 (3)</td>
<td>385</td>
</tr>
<tr>
<td>Congenital hypertrophy of retinal pigment epithelium</td>
<td>2 (3)</td>
<td>18; 19 (18-19)</td>
</tr>
<tr>
<td>Idiopathic choroiditis</td>
<td>2 (3)</td>
<td>25</td>
</tr>
<tr>
<td>Retinopathy of prematurity</td>
<td>2 (3)</td>
<td>300; 300 (0-600)</td>
</tr>
<tr>
<td>Histoplasmosis</td>
<td>1 (1)</td>
<td>22</td>
</tr>
<tr>
<td><strong>Total</strong></td>
<td><strong>67</strong></td>
<td><strong>56</strong></td>
</tr>
</tbody>
</table>

Abbreviation: VPT, vasoproliferative retinal tumor.

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Figure 1. Primary retinal vasoproliferative tumor showing various features of intraretinal exudation (A-D and E), subretinal/intraretinal hemorrhage (A, B, D, E, and F), shallow subretinal fluid (A, B, D, and E), and retinal pigment epithelial alterations (A and C).
Figure 2. Secondary retinal vasoproliferative tumor with underlying conditions of retinitis pigmentosa (A), postretinal detachment surgery (B), retinopathy of prematurity (C), and pars planitis (D).

Figure 3. Primary retinal vasoproliferative tumor before (A) and after (B) cryotherapy.
VPTs, and in most cases, the underlying condition had been established prior to the detection of the secondary VPT. Important similar features of primary vs secondary VPTs included tumor location in the inferotemporal quadrant, tumor location between the equator and ora serrata, tumor thickness, and presence of remote macular findings (Table 1 and Table 2).

A notable finding in this analysis was the prevalence of a visually damaging effect of peripheral VPT on macular function. Visual acuity at presentation was 20/60 or worse in 39% of cases, and in these cases, treatment was provided for tumor control and visual rehabilitation. Poor visual acuity with VPT has been previously recognized in published series of 103 eyes by Shields and associates. More other publications on this topic have been small case series or single-case reports, lacking sufficient data to evaluate visual outcome. In our current series of 295 eyes, both primary and secondary VPTs showed poor visual acuity of 20/200 or worse in 18% of patients from macular exudation, macular edema, epiretinal membrane, or retinal traction. This is an often overlooked cause of macular dysfunction and clinicians should be vigilant in examination of the peripheral fundus for conditions that can affect the macula, such as VPT.

In summary, VPT can occur as a primary or secondary tumor, each with potentially profound effects on the macula and ultimate visual outcome. Both types can have similar features and complications, but secondary VPTs are more likely to affect younger patients, both eyes, with multifocal lesions and overall poorer visual acuity. The diagnosis of VPT, particularly in a younger patient, should prompt a detailed ocular and systemic investigation for an underlying condition.

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