22 Vascular Abnormalities

22.1 Idiopathic Juxtafoveolar Retinal Telangiectasis

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Core Messages
- Retinal telangiectasis is a group of rare retinal vascular anomalies affecting the retinal capillaries
- Characteristically irregular dilation, leakage and edema occur in the macula, rarely combined with vascular changes in the retinal periphery
- Telangiectasis may develop unilaterally or bilaterally and is commonly characterized by a slow decrease in visual acuity in adulthood
- The long-term prognosis for reading vision is usually good
- The pathogenesis of these changes is unknown

22.1.1 History

Idiopathic juxtafoveolar retinal telangiectases (IJRT) were classified by Gass and Oyakawa in 1982 using biomicroscopic and fluorescein angiographic data [6]. This classification was updated by Gass and Blodi in 1993 [5], who subdivided IJRT into three groups (Table 22.1.1). Group 1 IJRT are unilateral in most cases and characterized by dilated retinal capillaries and abnormal leakage leading to an easily visible exudation. Group 2 IJRT are mostly bilateral and characterized by late staining on fluorescein angiography with minimal exudation. Later in the disease process retinal pigment epithelial proliferation or secondary subretinal neovascularization may develop. In Group 3 changes are based on bilateral capillary occlusion. These changes lead to easily visible telangiectasis, parafoveolar capillary occlusion and minimal exudation.

Table 22.1.1. Summary of findings in idiopathic juxtafoveolar retinal telangiectasis [5]

<table>
<thead>
<tr>
<th>Group</th>
<th>No.</th>
<th>Localization</th>
<th>Visual acuity</th>
<th>Mean age (years)</th>
<th>Gender</th>
<th>Characteristic</th>
</tr>
</thead>
<tbody>
<tr>
<td>1A</td>
<td>31</td>
<td>Unilateral (97%)</td>
<td>20/40</td>
<td>37</td>
<td>Male</td>
<td>Leakage</td>
</tr>
<tr>
<td>1B</td>
<td>8</td>
<td>Unilateral (88%)</td>
<td>20/20</td>
<td>42</td>
<td>Male</td>
<td>Leakage</td>
</tr>
<tr>
<td>2A</td>
<td>92</td>
<td>Bilateral (98%)</td>
<td>20/40</td>
<td>55</td>
<td>No specificity</td>
<td>Diffusion</td>
</tr>
<tr>
<td>2B</td>
<td>2</td>
<td>Bilateral (100%)</td>
<td>20/70</td>
<td>11</td>
<td>Diffusion</td>
<td></td>
</tr>
<tr>
<td>3A</td>
<td>3</td>
<td>Bilateral (100%)</td>
<td>20/25</td>
<td>53</td>
<td>Occlusion</td>
<td></td>
</tr>
<tr>
<td>3B</td>
<td>4</td>
<td>Bilateral (100%)</td>
<td>20/50</td>
<td>42</td>
<td>Occlusion</td>
<td></td>
</tr>
</tbody>
</table>

22.1.2 Clinical Course of the Disease

Essentials
- Group 1: Unilateral telangiectasis with abnormal leakage of capillaries
  - Group 1A: Visible and exudative idiopathic juxtafoveolar retinal telangiectasis with lipid exudates, size > 1 disk diameter
  - Group 1B: Visible and exudative focal idiopathic juxtafoveolar retinal telangiectasis, size < 1 disk diameter
- Group 2: Bilateral idiopathic juxtafoveolar retinal telangiectasis
  - Group 2A: Nonexudative bilateral idiopathic juxtafoveolar retinal telangiectasis
  - Group 2B: Juvenile occult familial idiopathic juxtafoveolar retinal telangiectasis
- Group 3: Occlusive idiopathic juxtafoveolar retinal telangiectasis
  - Group 3A: Occlusive idiopathic juxtafoveolar retinal telangiectasis without central nervous system vasculopathy
  - Group 3B: Occlusive idiopathic juxtafoveolar retinal telangiectasis with central nervous system vasculopathy
In **Group 1A** primarily male patients are affected. Commonly this group is characterized by a unilateral development of retinal telangiectasis and in most cases the vascular changes are easily visible. Patients with unilateral IJRT may be asymptomatic or may experience a mild reduction of visual acuity. Biomicroscopic findings are prominent telangiectatic retinal capillaries in the temporal half of the fovea with a 2 DD involvement of the macula, mostly associated with surrounding intraretinal lipid exudates (Fig. 22.1.1a).

**Group 1B** telangiectasis differs primarily in the size of lesion from Group 1A. The extension of telangiectatic vessels is only 1 disk diameter temporal of the fovea and the changes are mostly not associated with lipid exudates (Fig. 22.1.2). Male predilection, unilaterality and biomicroscopic and fluorescein angiographic features are nearly the same. It may be a mild form of Group 1A telangiectasis [5].

Patients in **Group 2A** IJRT have the most common form of idiopathic juxtafoveal retinal telangiectas-