Acute Zonal Occult Outer Retinopathy (AZOOR)

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Core Messages

- Acute zonal occult retinopathy is a syndrome first described by Gass [3, 7], who introduced the acronym AZOOR, which summarizes the typical clinical characteristics of a spectrum of retinal disorders:
- *Acute:* rapid loss of visual function in one or both eyes with photopsias in the area of visual field
- *Zonal:* visual loss occurring in one or more retinal regions with or without concomitant blind spot enlargement
- *Occult:* minimal initial ophthalmoscopic changes or absence of funduscopically visible alterations in the retinal area corresponding to the visual field loss
- *Outer:* affecting primarily the photoreceptor and retinal pigment-epithelial (RPE) layer with abnormal responses on electroretinographic (ERG) testing. Cones tend to be more affected than rods
- *Retinopathy*
- Similar changes described in patients with *multiple evanescent white dot syndrome* (MEWDS), *acute idiopathic blind spot enlargement syndrome* (AIBSES), *multifocal choroiditis and panuveitis* (MCP), and *acute macular neuroretinopathy* (AMN). Therefore, it has been suggested that these entities are not separate diseases but an overlapping spectrum of a single disorder (*AZOOR complex*)
- *Acute annular outer retinopathy,* which may be a variant of AZOOR

4.1 Aetiology

The aetiology of AZOOR is still unclear, but it is presumed to be of autoimmune inflammatory origin. Evidence for autoantibodies directed against retina-specific proteins is still lacking [10]. Gass speculated that AZOOR may originate from a viral infection latent in a region of the outer retina which becomes activated, resulting in acute retinal dysfunction and potentially death of the retinal receptors with no immediate effect on funduscopic retinal appearance.

4.2 Clinical Findings

Photopsia and sudden visual field loss in one or both eyes typically in young, Caucasian (90 %), myopic women (f:m = 3:1) in their early thirties are characteristic clinical symptoms and findings in the initial phase of the disease.

4.2.1 Photopsia

The visual sensations in the early phase are described by almost 90 % of patients as multicoloured and associated with shimmering or amoeboid micro-movements in the area of visual field loss. They may be

exacerbated by bright light, stress, fatigue and exercise. These photopsias tend to persist.

4.2.2 Loss of Visual Field

Defects in the visual field are most commonly noted in the superior and temporal quadrants and are usually asymmetric (Fig. 4.1). However, any portion or almost the entire visual field may be involved. They almost always include the blind spot (90 %), which is often enlarged. The size of defects often increases within days or weeks before stabilizing. Visual field testing is probably the best parameter to monitor the disease and should be repeated regularly. In the long-term follow-up study of Gass reviewing 51 patients for a minimum of 3 years, visual field changes stabilized within 6 months in 78 % of patients, progressed in 4 %, and partially improved in 20 % [7]. Over time, the visual field defect can enlarge and can move peripherally or centrally.

Patterns of visual field loss caused by AZOOR in descending order of frequency are blind-spot enlargement, ring scotomas, hemianopic scotomas, 360-degree concentric contraction, arcuatelike scotomas, and multiple isolated scotomas [7].

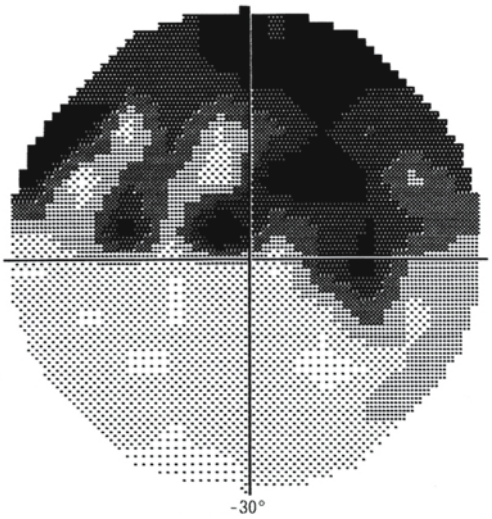

Fig. 4.1. Zonal visual field loss in the upper hemisphere including the blind spot (same eye as in all other figures)

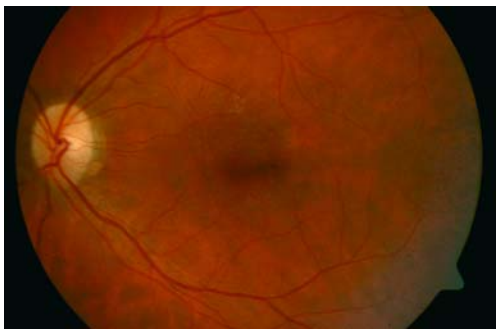

Fig. 4.2. Normal fundus appearance with subtle RPE changes in the macular area (same eye as in all other figures)

4.2.3 Fundus Changes

Early in the disease subtle pigment epithelial (RPE) changes may be noted on funduscopy (Fig. 4.2). However, in many patients no visible alterations are seen (hence the term "occult"), which may give rise to misdiagnoses and unnecessary neurologic and neuroradiologic work-up.

In later stages of the disease the visual field defects correspond to areas of visible pigmentary alterations. In areas of atrophy, retinal vessels may become narrowed. Migration of RPE can mimic the bone spicule appearance of retinitis pigmentosa (RP). Segmental perivenous sheathing may also occur.