Paracentral Acute Middle Maculopathy

*Paracentral acute middle maculopathy (PAMM)* refers to macular lesions with changes in the inner nuclear layer on SD-OCT. The primary etiology in PAMM may be ischemia of the deep capillary system, which is responsible for blood supply to the middle retina.

The typical presentation is acute onset of diminished central visual acuity (although Snellen measurement of 20/20 is possible) or paracentral scotoma. Ophthalmoscopically, the lesions may appear only as subtle parafoveal gray-white spots or wedges. Compared with cotton-wool spots, the retinal whitening associated with PAMM lesions is more distinct, duller gray-white, less opaque, and deeper in the retina; also, it is not distributed along the NFL. However, these lesions are evanescent and may resolve before clinical examination takes place. In such cases, the characteristic hyperreflective bands on SD-OCT should still be detectable (Fig 6-24). Over time, PAMM lesions typically resolve with thinning of the inner nuclear layer, resulting in persistent paracentral scotomata.

PAMM is primarily a disease of retinal ischemia and often seen in association with retinal vascular occlusion. Evaluation in suspected cases includes imaging and systemic workup for cardiovascular risk factors and sickle cell disease.

Arterial Macroaneurysms

Retinal arterial macroaneurysms are acquired ectasias of the first 3 orders of retinal arterioles. Large macroaneurysms can actually traverse the full thickness of the retina. Vision loss may occur from embolic or thrombotic occlusion of the end arteriole (white infarct) or from hemorrhage in any retinal layer. Other retinal findings may include capillary telangiectasia and remodeling, as well as retinal edema and exudate involving the macula (Fig 6-25). Often, there are multiple arterial macroaneurysms, although only 10% of cases are bilateral. Arterial macroaneurysms are associated with systemic arterial hypertension in approximately two-thirds of cases and may occur in the area of previous vascular occlusions. Systemic blood pressure should be measured at the time of diagnosis, and the patient should be referred for further evaluation.

Typically, the macroaneurysm closes and scleroses spontaneously, with accompanying resorption of related hemorrhage. Reopening of the macroaneurysm and rebleeding are rare. Thus, initial management is usually observation. Laser photocoagulation treatment may be considered if increasing edema in the macula threatens central vision. In most instances, closure can be achieved with moderate-intensity laser treatment of the retina, performed immediately adjacent to the macroaneurysm, using 2–3 rows of large-spot-size (200–500 μm) applications. Some specialists prefer direct treatment. Caution

Figure 6-24  Paracentral acute middle maculopathy (PAMM). A 57-year-old woman presented with acute visual defects that she described as “lacy patterns” in her vision. A, Fundus photograph shows ill-defined grayish lesions in the macula, corresponding to intraretinal opacification. B, Fundus autofluorescence highlights the blocking defect of the perivenular retinal opacification. C, SD-OCT shows hyperreflectivity of the inner nuclear layer, more patchy nasally and more continuous temporally. These clinical and imaging findings are consistent with PAMM. (Courtesy of Amani Fawzi, MD.)

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According to the ICROP, an eye is classified on the basis of the most advanced disease noted. However, documentation should reflect all affected zones and stages observed, including their relative extent.

**ACTIVITY 8-1** Interactive schematic for type 1 ROP.
*Developed by Franco M. Recchia, MD.*


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Figure 8-8  Aggressive ROP (A-ROP). Fundus photograph shows prominent plus disease and ill-defined retinopathy in zone I, accompanied by blot hemorrhages. (Courtesy of Franco M. Recchia, MD.)

Pathophysiology of ROP

A link between excessively exuberant perinatal oxygen supplementation and severe ROP was well recognized by the 1950s. After substantial reductions in oxygen use in neonatal intensive care units (NICUs), the incidence of ROP decreased dramatically. However, many infants experienced adverse neurologic outcomes as an unintended consequence of that oxygen restriction, and infant death rates rose. Once oxygen was again used more liberally, neurologic outcomes and survival improved, with the consequence of a resurgence of ROP.

Table 8-2 Common Terms Used in Clinical Trials to Describe Acute Retinopathy of Prematurity (ROP)

<table>
<thead>
<tr>
<th>Threshold disease (all 3 features must be present)</th>
<th>Prethreshold disease</th>
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<tr>
<td>Extraretinal neovascularization (stage 3 disease): EITHER 5 contiguous clock-hours OR 8 cumulative clock-hours Retinal vessels ending within zone I or zone II Plus disease</td>
<td>All zone I and zone II changes, except zone II stage 1 and zone II stage 2 without plus disease, that do not meet threshold treatment criteria; subdivided into type 1 and type 2 disease</td>
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<tr>
<td>Type 1 ROP Zone I, any stage ROP with plus disease, or Zone I, stage 3 ROP without plus disease, or Zone II, stage 2 or 3 ROP with plus disease</td>
<td>Type 2 ROP Zone I, stage 1 or 2 ROP without plus disease, or Zone II, stage 3 ROP without plus disease</td>
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