Peri-Papillary Atrophy

Maryke Neiberg, OD

Author's Bio

Dr. Maryke Neiberg is an associate professor and full-time faculty at Western University College of Optometry in Pomona, California. Any opinions expressed in this article are her own and she has no financial interest to disclose.

As optometrists, the optic nerve and rim tissue are of great interest to us. We look very carefully at the disc margins, color, and cup-to-disk ratio. We certainly look at the nerve fiber layer, and we may even pay special attention to the tissue directly surrounding the optic nerve. What can this area tell us?

To answer this question, we would have to know a little about the unique anatomy of this area. This area is significant in terms of its vascular supply. It shares its vascular supply with that of the prelaminar, laminar and retrolaminar optic nerve. This section of the optic nerve and the peri-papillary area is supplied by the circle of Zinn-Haller, which represents anastomosis of the short posterior cilliary arteries.[1] Usually, this area appears uniform in pigmentation with a clear demarcation between retina and optic nerve.

In some cases, the optic nerve is surrounded by an area of hyper and hypopigmentation that may encircle the disc margin, commonly more visible in the temporal disc area.[2] This pigment disturbance or mottling represents chorioretinal atrophy around the optic disc. This chorioretinal atrophy is known as peri-papillary or para-papillary atrophy (PPA), and is a relatively common finding. [3] Fluorescein or indocyanogreen angiopathy show characteristic filling patterns that can be used to further differentiate the peripapillary atrophy.

The clinical description of PPA should be distinguished from the physiological grey crescent that surrounds the optic nerve. The grey crescent represents a localized deposition of pigmentation demarcating the edge of the optic disc.[7] Other crescents that can be noted around the optic nerve include the myopic crescent, which is a white sharply demarcated crescent to the temporal side of the optic disc, presents bilaterally and is generally associated with axial myopia. This type of crescent does not show pigment mottling that is typically associated with peripapillary atrophy . The choroid is absent in this area, the retina is transparent and the sclera appears as a white crescent. In pathological myopia, the atrophic crescent may increase in extent.

Contrasting with a scleral crescent, a choroidal crescent represents absent pigment epithelium in a temporal crescent at the disc. The bare choroid is present and its vessels are visible. [3]

In addition to the physiological grey crescent, tilted or mal-inserted nerves could also prompt a diagnosis of PPA. Mal-insertion may add to the glaucomatous appearance of the nerve.[8, 9] It is well known that tilted discs are associated with superior temporal visual field defects, but that other types of visual field defects could also accompany tilted discs.[10]

The glaucomatous looking nerve with visual field defects could present a diagnostic dilemma for the clinician, but repeated visual field evaluation will show a stable visual field with non-progression when the field defects are due to the tilted disc and not glaucomatous. Such obvious and frank mal-insertion affords the unique opportunity to observe the vascular circle of Zinn-Haller with fluorescein angiography in vivo.[11, 12]

Mal-insertion might present so subtly that it remains subclinical.[3] Histologically, however, there are three distinct types of PPA identifiable. The first two types are expressions of congenital misalignment and malposition. The first type is due to simple misalignment of the choroid and the retinal pigment epithelium (RPE)-this gives rise also to a choroidal crescent. The scleral stretching that accompanies the misalignment may resemble age related atrophic PPA.[3] The second type is due to the mal-positioning of the embryonic fold between the RPE and the outer retina.[3]

The third histologically identifiable type of PPA is due to age related atrophy of the RPE and the rods. This presentation may be observed on a continuum of complete absence of the RPE, to loss of pigmentation of the RPE.[3] Clinically, therefore, description of peri-papillary atrophy includes, and what can histologically be described as true tissue atrophy.[3]

Normal aging and vascular factors

The choriocapillaris plays a vital role in maintaining the RPE. The decreased coverage of Bruch's membrane by the choriocapillaris leads to RPE degeneration. The degree, to which the choriocapillaris is affected, is expressed by the extent to which the degeneration or the atrophy of the RPE is expressed. [13] The basal lamina thickens with age while the RPE degenerates and becomes atrophic. [13] The aging eye shows evidence of choroidal thinning, often with marked tessellation of the fundus. This type of degeneration of the choroid is more commonly associated with zone beta PPA.[4, 6]

In addition to the blood supply from the choriocapillaris, the position of the central retinal artery trunk is thought to play a role in the development of PPA.[6, 14] The quadrant that contains the vascular trunk often shows less peri-papillary atrophy. [14] Although vascular support from the central retinal trunk to the peri-papillary area has not been demonstrated, the position of the trunk is thought to provide mechanical support to the lamina cribrosa.[15] Similarly, the cilio-retinal arteries may on occasion support the pre-laminar region of the optic nerve, but the presence or position of the cilio-retinal artery does not affect peri-papillary atrophy.[6, 14, 16]

The macula and periphery of normally aging eyes present with degeneration of the Bruch's membrane complex. The atrophy from PPA is similar to these degenerative changes. When PPA is associated with macular degeneration, the risk for developing glaucoma is increased.[4] The reverse seems to hold true too. The optic nerves of elderly patients with glaucoma seem to have an accelerated risk for developing PPA.[4] The zone of peri-papillary retinal pigment epithelium atrophy becomes significantly wider after the age of 75 and is mostly associated with a loss of rods, not cones, at the termination of Bruch's membrane. [13]

PPA is generally associated with congenital events and that additional and progressive atrophy is known to occur with aging. [13] Two distinct zones can be observed surrounding the optic nerve. Each of these zones has diagnostic and clinical significance. Jonas et al., named the area closest to the disc where the sclera and choroidal vessels are visible, the beta zone (or zone beta).[6] The smaller zone beta is characterized by the termination of photoreceptor layer and retinal pigment epithelium layers.[3]

Zone beta is always closer to the optic nerve; it extends from the peri-papillary scleral ring to the edge of zone alpha.[6] They used the designation of zone alpha to describe the atrophic area of irregular pigment mottling, consisting of areas of hypo-and hyperpigmentation which *leads* into the retina.[6, 3] Zone alpha represents photoreceptor and underlying pigment epithelium layer disruption.[3] Angiography with sodium fluorescein or indocyanine green shows typical findings:

Zone beta shows no choroidal filling, indicative of the absence of the vascular choriocapilaris.[4, 5, 6] Zone alpha shows decreased choroidal filling, and this finding is typical in open angle glaucoma. [4, 5]

Zone alpha is more common than zone beta. A study of Chinese put the prevalence of zone alpha at 70% and that of zone beta at 20% of elderly subjects.[17] The visual field defect caused by zone alpha is relative, while that of zone beta is absolute.[3,18]

The decrease of vascular supply by the choriocapillaris is a significant factor in the genesis of PPA. Conversely, the damaging effect of hypertension was demonstrated in an animal study involving rhesus monkeys. The study showed that chronic arterial hypertension caused optic nerve head damage and atrophic changes in the temporal peri-papillary choroid.[19] This makes a cogent case for the control of chronic arterial hypertension in our human patients.

Myopia

Peri-papillary atrophy is commonly observed together with high myopia, estimated to be present in about 20% of cases.[20] In highly myopic eyes, the scleral crescent may form a complete ring around the optic nerve. This additional scleral ring may accompany the alpha and beta zones and is indicative of the absence of Bruch's membrane. [21, 22] A temporal myopic crescent is present in about a third of cases of patients with high myopia. [20, 21] The congenital cause and presentation of peri-papillary atrophy is not always clinically appreciable, but would certainly be histologically identifiable.

High myopia and beta-peri-papillary atrophy occur together, but are not inherited from the same genes.[3] They reflect the coinheritance of genes specific for beta peri-papillary atrophy and myopia. [3] In addition to the congenital programing of beta-peri-papillary atrophy, environmental factors contribute to the total visible atrophy.[3] Therefore, if the temporal crescent increases in size in a patient with physiologic myopia and glaucoma, it could be interpreted as a marker that the glaucoma is progressing.[23]

In general, disc hemorrhage and open angle glaucoma is known to be increased in the elderly myopic population. [24] Optic pits are also associated with beta peri-papillary atrophy in the same population. [24]

Glaucoma

Peripapillary atrophy is not a typical feature of the eye with **ocular hypertension**. It seems more resilient in terms of developing PPA.[25, 28]

In the early stages of **open angle glaucoma**, the volume and area of temporal peri-papillary atrophy are closely related to the pathogenesis of the disease. [25] The presence of PPA is not a clinically useful parameter to diagnose early open-angle glaucoma nor to differentiate it from normal-tension glaucoma, but it is helpful to observe and follow when some of the other parameters are not available.[6, 25]

The extent and development of the peri-papillary atrophy is known to be related to the severity of the glaucomatous optic nerve damage and of the visual field defects. The extent and development of the peri-papillary atrophy is useful for the assessment of progression of damage of the disk. [21, 27, 28] A large zone beta may encircle the disc and is known as "halo glaucomatosus". [6]

If the PPA continues to deteriorate as measured by visual field, despite good control of intraocular pressure, this more likely is representative of the patient's vascular risk factors for progression of glaucoma.[15] In patients with glaucoma, peri-papillary focal arteriolar narrowing appears to be related to the severity of the glaucoma and can be associated with a visual field defect in the corresponding hemifield.[25]

Ocular Coherence Tomography is routinely used to evaluate the nerve fiber layer in patients with glaucoma. When significant peri-papillary atrophy is present, population derived normative age data should not be used in analysis, as this might lead to an overestimation of the glaucomatous damage.[29] Loss of the characteristic double hump pattern is seen, and the TSNIT graph shows irregular high spikes that make the pattern difficult to reliably interpret. [30]

Spectral domain OCT is thought to be superior in analysis, showing less of the irregular measurement spikes seen with time domain OCT. [31] A recent study of PPA using spectral domain OCT showed that retinal thickness in patients with peri-papillary atrophy is thinner compared to patients without, but that the there was no statistical difference in the thickness of the respective nerve fiber layers.[32]

Most of the peri-papillary atrophy associated with **normal-tension glaucoma** occurs inferior to the optic disc.[33] PPA significantly reflects visual field (functional) and optic nerve (structural) damage in normal tension glaucoma. The location of the visual field defects correspond significantly with the location of the PPA, and the topography of the nerve corresponds with zone beta. [34] When beta zone PPA is present in normal tension glaucoma, it is certainly indicative of progression of visual loss. [12]

Focal rim notching is known to commonly precede a disk hemorrhage, with the hemorrhage usually occurring adjacent to the notch. [35] Peri-papillary atrophy precedes disk hemorrhage in 80% of cases. This type of disk hemorrhage is most commonly seen in females over the age of 60 with associated PPA.[36] This association is very significant, occurring irrespective of small neuroretinal rim area or volume. [35]

Acute angle closure does not show a direct relationship with PPA. The peri-papillary atrophic area does not enlarge despite an increase in the optic cup after angle closure.[36] This differentiates the mechanisms for development of optic disk damage between acute angle closure and primary open angle glaucoma. [37]

While the effect of the choriocapillaris on the peri-papillary tissue in patients with glaucoma is undeniable, PPA does not progress after anterior ischemic optic neuropathy. [6] The disk itself shows defective filling, supporting the difference in pathologic mechanism.[38] Pseudoexfoliation without glaucoma is also not directly associated with PPA. [14]

Ocular Disease

Several ocular diseases are associated with peri-papillary atrophy. These diseases are listed in Table 1. The etiology of PPA cannot be visually determined, and therefore, the etiology is clinically indistinguishable. [3]

Table 1. Ocular Diseases and Conditions that are Associated with Peripapillary Atrophy [39- 50]
Stargardt's Disease
Helicoid Peripapillary Atrophy (Sveinsson Chorioretinal Atrophy)
Vogt-Koyanagi-Harada
Idiopathic Multifocal Choroiditis
Multiple Evanescent White Dot Syndrome (MEWDS)
Serpiginous Choroiditis
Toxoplasmosis
Histoplasmosis
X-linked Retinitis Pigmentosa
Sympathetic Ophthalmia
Angioid Streaks
Glaucoma
Autosomal Dominant Optic Atrophy OPA 1

Conclusion

Peri-papilary atrophy is a fairly common condition that is highly associated with aging. It may also be genetically expressed in conjunction with myopia and may also be seen in conjunction with optic pits or tilted and mal-inserted disks. The mal-insertion may be clinically visible or subclinical.

There are several ocular and systemic diseases that are associated with PPA. The choriocapillaris supplies the perineural area and the condition, extent and progression of the peri-papillary atrophy directly reflects the circulatory or vascular risk factors of the patient.

The progression of peri-papillary atrophy is very closely associated with open angle glaucoma and normal tension glaucoma. Ocular hypertension, primary angle closure and pseudoexfoliation are not associated with the presence or progression of peri-papillary atrophy.

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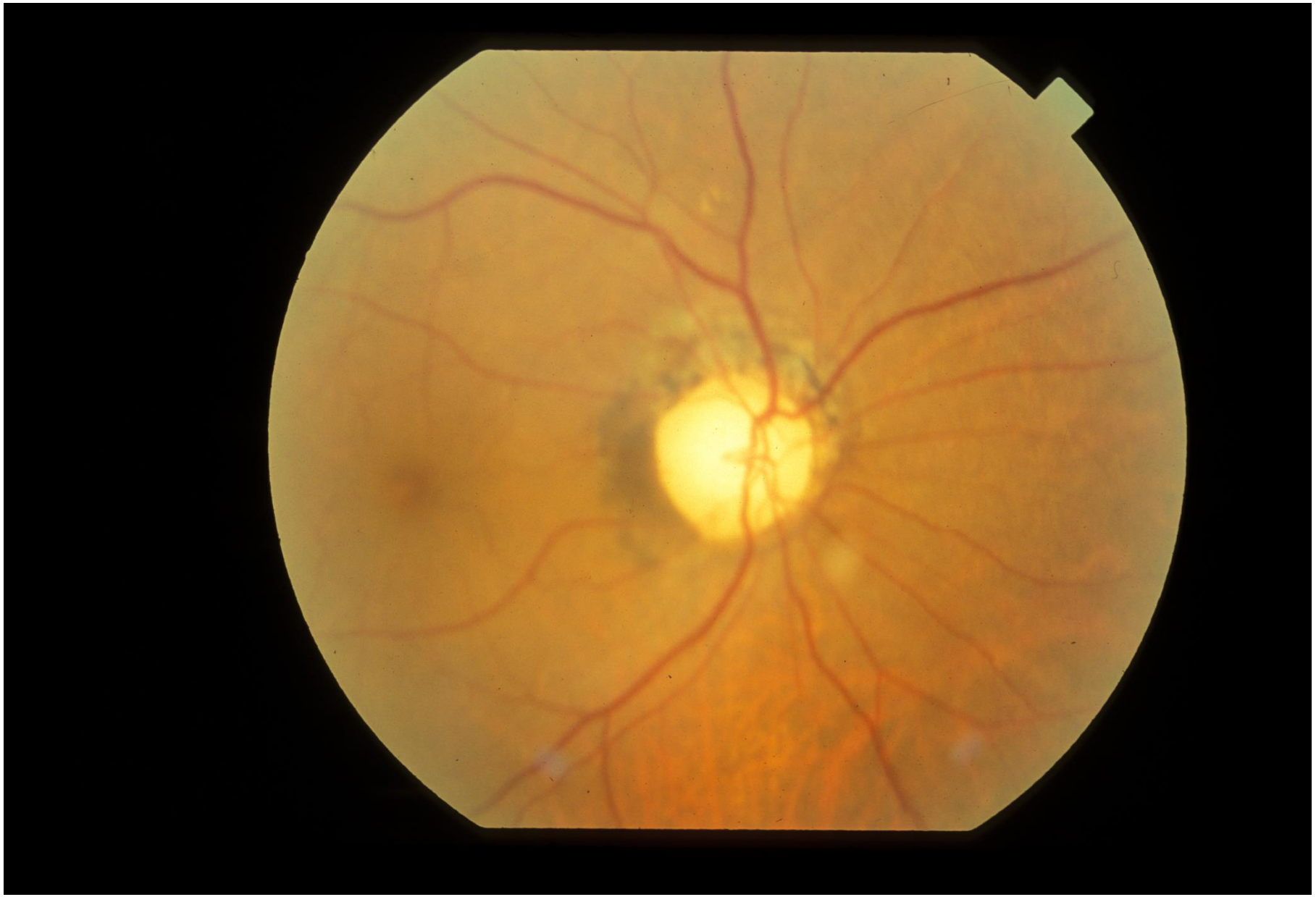
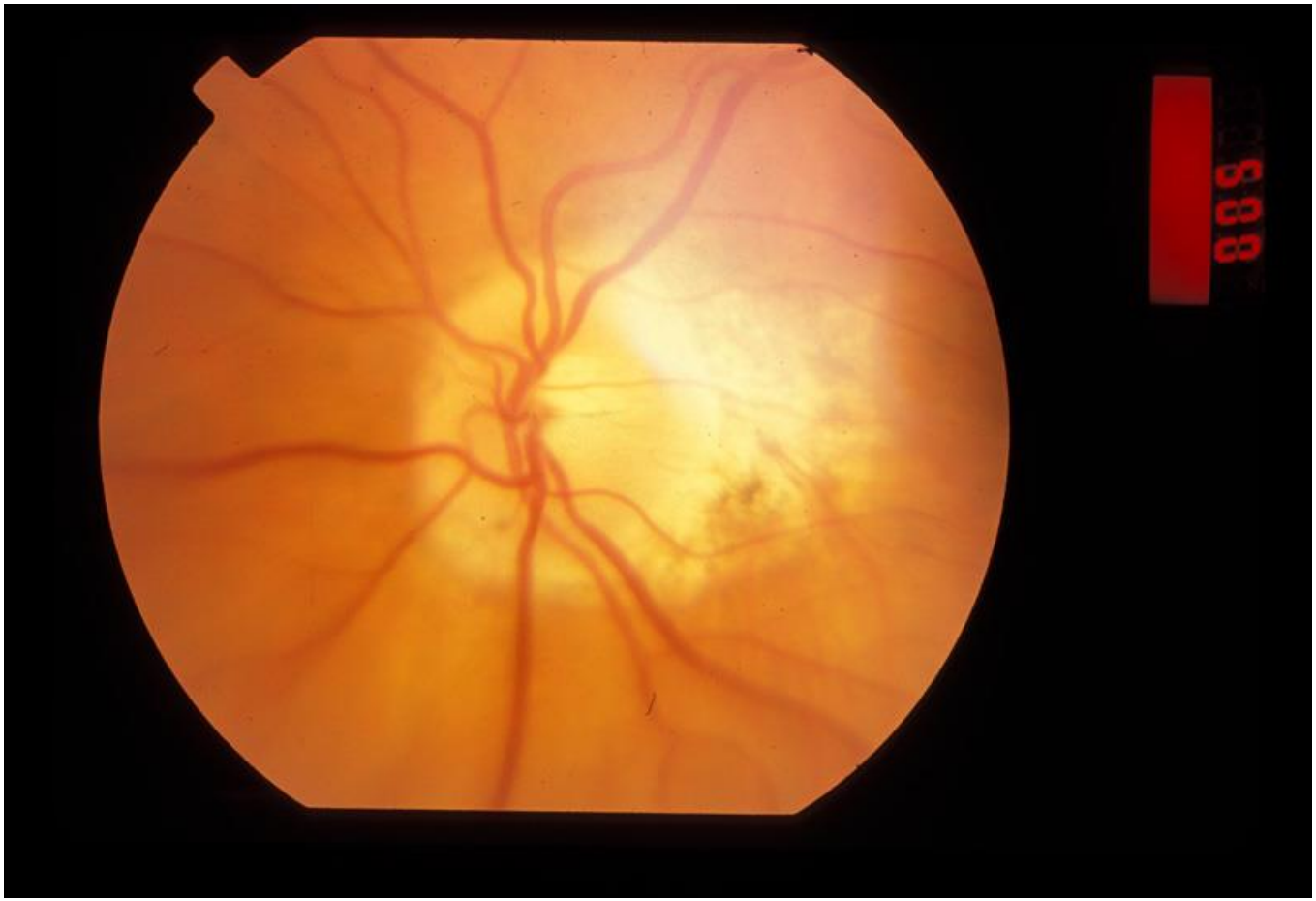
Images for Peripapillary Atrophy

Peripapillary atrophy, glaucoma and attenuated blood vessels heighten concern over this patient’s vascular risk factors



Significant temporal peripapillary atrophy can be seen at the temporal disc margin in this patient.



Presentation of helicoid peripapillary atrophy around the optic disc of this patient