Nonglaucomatous Optic Atrophy

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KEYWORDS

• Optic atrophy • Optic disc • Optic neuropathy • Pallor

DEFINITION

In clinical terms, optic atrophy occurs when the optic disc is thought to be less pink or paler than normal (**Fig. 1**). Thus, optic atrophy is not a diagnosis but simply an ophthalmoscopic sign similar to finding upper extremity weakness; the cause of the weakness must still be determined. Unfortunately, the term ''optic disc pallor'' is often used synonymously with optic atrophy. A wide range of ''normal'' optic disc color exists, and thus it can be difficult to be sure if optic atrophy is present. A normal optic disc is often pinker on the nasal side, with relative temporal pallor. The central cup of the disc appears pale because the white lamina cribrosa is visible. Optic discs in patients with axial myopia may appear paler. Furthermore, media opacities such as cataract can impart more color to the disc, and pseudophakia can result in a paler disc appearance. Thus, optic disc pallor should not be used synonymously with optic atrophy. It is essential to compare an individual's optic discs, because they will usually have the same color. If true atrophy (damage) exists, it should be accompanied by visual loss (acuity and/or peripheral vision), decreased color perception (if acuity is compromised), and a relative afferent pupillary defect unless the damage is symmetric. Ophthalmoscopic evaluation of the peripapillary retina may reveal diffuse or segmental nerve fiber layer loss. New technology such as optical coherence tomography can also be helpful in demonstrating axonal loss. Retinal vascular attenuation often occurs after anterior ischemic optic neuropathy (AION).[1] Comparison of vessels from eye to eye helps to confirm this finding and suggests previous ION.

Pathologically, optic atrophy is shrinkage of the optic nerve caused by degeneration of its axons. The axons may degenerate because of damage from anywhere in the ganglion cells in the retina to the point of axonal synapse in the lateral geniculate

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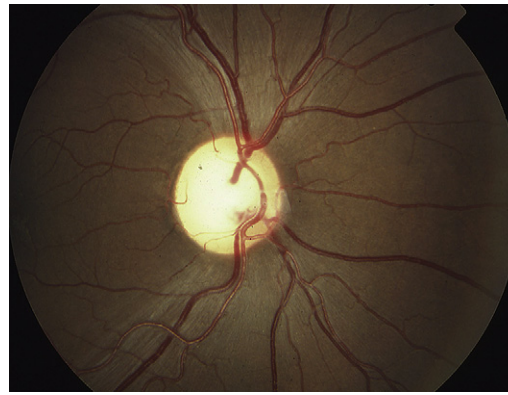

Fig. 1. Nonspecific diffuse optic atrophy.

body. The clinically apparent increase in pallor of the optic disc is not completely understood.[2] Loss of axons with resultant decreased blood supply and gliosis have been proposed causes.[3] Quigley and Anderson[4] proposed that loss of axons affects the normal light transmission and diffusion amongst capillaries and that the light is instead reflected by the remaining white glial tissue. These investigators did not find evidence of astroglial proliferation in atrophic optic nerves. Several investigators have used laser Doppler velocimetry to study optic disc blood flow in patients with atrophy[5,6] and have found diminution of blood flow in such optic discs, but of mild nature. Thus, questions on the exact cause of the pallor observed in optic atrophy still exist.[2]

Rarely, trans-synaptic degeneration from damage to the retrogeniculate visual pathway results in optic atrophy, which occurs primarily in the immature brain (in utero or early infancy).[7] Clinically apparent transsynaptic degeneration does not typically occur in human adults, although it has been demonstrated pathologically in some patients.[8]

DIFFERENTIAL DIAGNOSIS

Anything that damages the retinal ganglion cells or their axons will result in optic atrophy. Most commonly, the atrophy is diffuse and nonspecific, although exceptions do exist (see **Fig. 1**). It is beyond the scope of this article to completely discuss glaucomatous optic atrophy. However, it is important to emphasize the differences between glaucomatous and nonglaucomatous optic atrophy. The hallmark of glaucomatous optic atrophy is a progression in the size of the optic disc cup. The cupping is typically accentuated vertically, thus causing a more vertical than horizontal cup (**Fig. 2**). Focal notching of the disc is also characteristic of glaucoma. Nevertheless, conditions other than glaucoma (eg, compression, ischemia) have been reported to cause an increase in cup-to-disc ratio, but this is usually mild in relation to degree of optic nerve dysfunction.[9,10] Trobe and colleagues[11] had 5 ophthalmologists review 163 fundus stereophotographs of 9 entities as "unknowns." Glaucoma, central retinal artery occlusion, and ION were diagnosed by at least one observer with accuracy greater than 80%. Vertical elongation of the cup was the most important factor in differentiating glaucoma from other conditions. Another important differentiating characteristic was the rim pallor (**Fig. 3**). Thus, even in the presence of definite glaucoma,