Optic nerve head drusen associated with abnormally small optic discs

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Summary

Using Littmann’s method for correcting the magnification of central fundus photographs we evaluated the absolute optic disc size in 26 eyes with visible optic nerve head drusen. The optic nerve head area in these eyes (1.79 ± 0.50 mm²) was significantly smaller (p<0.001) than normal standard values previously determined (2.89 ± 0.76 mm²). The drusen were most commonly located and most densely packed at the upper and lower optic disc border. The coefficients of variation of the method’s reproducibility were 0.06 for intraobserver and 0.11 for interobserver determination.

The abnormally small optic disc indicating an abnormally small optic nerve scleral canal may inhibit by mechanical compression the axonal flow within the optic nerve fibers. This may ultimately lead to drusen formation. Pseudoneuritis also associated with an abnormally small optic disc may be a preciser of acquired optic nerve head drusen.

Introduction

Since their first description by Müller [12] in 1858 the pathogenesis of optic nerve head drusen is still not yet completely cleared. Tso [19] concluded from his electronmicroscopic and Seitz [17] from his light microscopic studies that optic disc drusen are related to axonal degeneration of optic nerve fibers. Spencer [18] in his Jackson Memorial Lecture as well as Rosenberg et al. [15] mentioned the coincidence of these ‘hyaline bodies’ with a congenitally small respectively dysplastic optic nerve head. Using only relative quantitative values Mullie and Sanders [13] measured indirectly the optic nerve head containing drusen and found some evidence for this theory.

It was this study’s purpose to evaluate the absolute dimensions of optic discs with drusen and to compare these measurements with previously determined standard values of normal optic nerve heads [4].

Patients and methods

Reviewing our hospital’s clinical and photographic files from 1980–1986 we found 26 optic nerve heads with visible drusen (7 men, 6 women, 13 right eyes, 13 left eyes). The diagnosis had been made ophthalmoscopically and confirmed by echography, autofluorescence in the course of fluorescence angiography, or orbital computer tomography. Patients with medical, otolaryngologic or neurologic findings concerning the optic nerve respectively

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patients suffering from optic nerve or retinal diseases leading to a prominence of the optic disc were excluded from the study.

The patients' ages ranged from 16 years to 52 years (mean 32.3 ± 11.9 years), their refraction from −4.5 diopters to +3.0 diopters (mean −0.53 ± 1.9 diopters).

The optic nerve head photographs (colour slides, 15 degree) were magnified 20 times, projected, and cup and disc borders plotted on white paper. Stereophotographs, colour contrast enhancement using red-free filters and changes in vessel course were helpful to outline the borders.

According to Littmann's method [10] the photographic magnification was evaluated taking into account the constant fundus camera and the individual ocular magnification factor. The validity of the Littmann formula has been confirmed by comparing intravital optic disc size measurements using Littmann's method and comparing direct measurements of optic nerve scleral canals in fresh enucleated, not fixed human donor eyes with [5].

Reproducibility. To detect the method's reproducibility we chose randomly photographs of 10 optic discs and reevaluated them ten times each. These measurements were carried out by J.J. and G.G. measuring these optic nerve heads.

Additionally we determined the drusen's locations, vessel abnormalities, existence of optic disc cupping, papillary or parapapillary hemorrhage and number of cilioretinal arteries.

Table 1. Dimensions of 26 unselected optic discs with drusen.

<table>
<thead>
<tr>
<th></th>
<th>Mean</th>
<th>Minimum</th>
<th>Maximum</th>
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<tbody>
<tr>
<td>Area (mm²)</td>
<td>1.79 ± 0.50</td>
<td>0.8</td>
<td>3.25</td>
</tr>
<tr>
<td>Minimal diameter (mm)</td>
<td>1.39 ± 0.19</td>
<td>0.97</td>
<td>1.87</td>
</tr>
<tr>
<td>Maximal diameter (mm)</td>
<td>1.66 ± 0.26</td>
<td>1.12</td>
<td>2.31</td>
</tr>
</tbody>
</table>

Table 2. Dimensions of 88 unselected normal human optic discs, high myopes < −8.00 diopters excluded (from Jonas JB, Gusek GCh, Guggenmoos-Holzmann I, Naumann GOH. Variability of the real dimensions of normal human optic discs).

<table>
<thead>
<tr>
<th></th>
<th>Mean</th>
<th>Minimum</th>
<th>Maximum</th>
</tr>
</thead>
<tbody>
<tr>
<td>Area (mm²)</td>
<td>2.89 ± 0.76</td>
<td>0.86</td>
<td>5.39</td>
</tr>
<tr>
<td>Minimal diameter (mm)</td>
<td>1.79 ± 0.25</td>
<td>0.85</td>
<td>2.43</td>
</tr>
<tr>
<td>Maximal diameter (mm)</td>
<td>2.05 ± 0.27</td>
<td>1.21</td>
<td>2.86</td>
</tr>
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</table>

Results

Optic disc area in eyes with drusen of the disc measured at average 1.79 ± 0.5 mm² (minimum 0.8 mm², maximum 3.25 mm²) (Table 1) and was significantly smaller (p<0.001) than previously determined standard values (2.89 ± 0.76 mm²) (Table 2) [4]. The minimal optic disc diameter ranged from 0.97 mm to 1.87 mm (mean 1.39 ± 0.19 mm), the maximal diameter from 1.12 mm to 2.31 mm (1.66 ± 0.25 mm). Also these values were significantly smaller (p<0.001) than those for normal human optic nerve heads [4] (Table 1, 2).

The drusen were most common and most densely packed at the upper and lower optic disc border followed by the disc's nasal side. At the temporal side they could generally be detected only when they were located circularly around the disc.

Vessel abnormalities were found in 5 eyes, but the central retinal artery was always nasal to the central retinal vein. There was a cilioretinal artery and a suggestion of an optic cup in 2 eyes. Concentric visual field constriction was found for 12 eyes (46.2%), enlargement of the blind spot in 13 eyes (50.0%) and arcuate scotomata in 10 eyes (38.5%). Hemorrhage within the optic nerve fiber layer at the disc's border was present twice. Method's reproducibility: Coefficients of variation for intraobserver evaluation were 0.06 and for interobserver determination 0.11.