The Glaucomatous Disc and Nerve Fibre Layer

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Introduction:
Increased intraocular pressure is one of the main risk factors for glaucomatous optic neuropathy. Since some subjects can tolerate a higher intraocular pressure before glaucomatous optic nerve fibre loss occurs than other individuals can, the presence of an elevated intraocular pressure alone is not sufficient for the diagnosis of glaucoma. Furthermore, epidemiological studies have shown that quite a number of patients with glaucomatous optic nerve damage have statistically normal intraocular pressure measurements. Other investigations have revealed that patients with evident glaucomatous optic nerve damage can have normal visual fields. Sommer and associates suggested that retinal nerve fibre layer abnormalities preceded visual field damage by four to six years.

The presence of glaucomatous visual field loss is, therefore, not necessary for the early diagnosis of glaucoma. The following is description of morphologic variables of the optic nerve head and retinal nerve fibre layer that may suggest early glaucomatous optic nerve damage.

A. Optic nerve head

1. Shape of neural rim:
In normal eyes, the neuroretinal rim shows a characteristic configuration. It is based on the vertically oval shape of the optic disc and the horizontally oval shape of the optic cup. The neuroretinal rim is usually broadest in the Inferior disc region, followed by the Superior disc region, the Nasal disc area, and finally the Temporal disc area (the ISNT rule), (Figures 1 and 2).
Fig. (1):
There neuroretinal rim is usually broadest in the Inferior disc region, followed by the Superior disc region, the Nasal disc area, and finally the Temporal disc region (the ISN'T rule).

Fig. (2):
The neural rim is almost equal all around. This should arouse the suspicion of possibility of glaucoma.

In eyes with modest glaucomatous damage, rim loss is found predominantly at the inferotemporal and superotemporal disc regions. In eyes with moderately advanced glaucomatous atrophy, the horizontal temporal disc region is the location with the most marked rim loss (Figure-3). In advanced glaucoma, the rim remnants are located mainly in the nasal disc sector, with a larger rim portion in the upper nasal region than in the lower nasal region. This sequence of disc sectors (inferior temporal - superior temporal - horizontal temporal - inferior nasal - superior nasal) correlates with the progression of visual field defects, with early perimetrical changes in the upper nasal quadrant of the visual field and a last island of vision in the inferior temporal part of the visual field in eyes with almost absolute glaucoma.

Fig. (3):
Moderately advanced damage of the neural rim with focal notching of the rim is seen at 6 o'clock. Field damage reflects the disc damage.
2. Optic disc hemorrhages:
Splinter-shaped or flame-shaped hemorrhages at the border of the optic disc (Figure-4) are a hallmark of glaucomatous optic nerve atrophy. Rarely or very rarely found in normal eyes, disc hemorrhages are detected in about 4% to 7% of eyes with glaucoma.
In two epidemiological studies, the frequency of disc hemorrhages in non-glaucomatous eyes was about 1%. In glaucomatous eyes, about two months after the initial bleeding, a localized defect of the retinal nerve fibre layer or a broadening of a localized retinal nerve fibre layer defect can be detected correlating with a circumscribed scotoma in the visual field. Since they are only rarely found in normal eyes; they usually indicate the presence of glaucomatous optic nerve damage, even if the visual field is unremarkable, and they suggest progression of glaucoma.
Glaucoma, however, is not the only optic nerve disease in which optic disc hemorrhages can be found. Therefore, they are as a single variable not at all sufficient to separate normal eyes and eyes with early glaucoma.

3. Parapapillary chorioretinal atrophy (PPA)
Ophthamoscopically, the parapapillary chorioretinal atrophy has been divided into a central beta zone and a peripheral alpha zone. The a zone is characterized by an irregular hypopigmentation and hyperpigmentation and intimated thinning of the chorioretinal tissue layer. On its outer side it is adjacent to the retina, and on its inner side it is in touch with a zone characterized by visible sclera and visible large choroidal vessels (β zone), or with the peripapillary scleral ring, respectively. Features of the β zone are marked atrophy of the retinal pigment epithelium and of the choriocapillaris, good visibility of the large choroidal vessels and the sclera, thinning of the chorioretinal tissues, and round bounds to the adjacent α zone on its peripheral side and to the peripapillary scleral ring on its central side.
If both zones are present, β zone is always closer to the optic disc than α zone.
In normal eyes, both α zone and β zone are largest and most frequently located in the temporal

Fig.(4):
Disc hemorrhages are detected in about 4% to 7% of eyes with glaucoma.
horizontal sector, followed by the inferior temporal area and the superior temporal region. (Figure-5) On the other hand in glaucomatous eye, the location of parapapillary atrophy (PPA) is spatially correlated with the neuroretinal rim loss in the intrapapillary region. (Figure-6)

**Fig.(5):**
In normal eyes, PPA is largest and most frequently located on temporal side then superior and inferior and are least prominent on the nasal side.

**4. Optic disc Cupping:**
Assessment of the size of the optic disc is important, since the small the optic disc should have small optic cup and the large disc should have large cup. (Figure-7) In ocular hypertensive subjects with small optic discs, small optic cups or low cup-to-disc ratios should therefore not prevent the diagnosis of glaucoma. also, large optic cups in large optic discs in ocular hypertensive subjects do not necessarily indicate glaucoma, since large optic discs physiologically have large optic cups.

**Fig.(6):**
The PPA atrophy has been divided into a central beta zone and a peripheral alpha zone. In glaucomatous eye, the location of PPA is spatially correlated with the neuroretinal rim loss.

**Fig.(7):**
Large optic discs usually have a large optic cup (a) while in small optic discs, cupping normally does not occur (b)
The optic disc area shows an inter-individual variability from about 0.80 - 6.00 mm² in a normal Caucasian population. Size of the optic disc depends on race, whites have relatively small optic discs, followed by Mexicans, Asians and Afro-Americans.

**Measuring the size of the disc:**
On slit lamp, the slit beam is widen to cover 1/3 of disc area, then the length of slit beam is adjusted so that it covers the disc from upper edge to lower edge. This length is measured from scale found on the slit lamp. Length is then multiplied by a correcting factor depending on the lens used for fundus examination. (see table).

**Correcting factor for the Volk lens:**

<table>
<thead>
<tr>
<th>Lens power</th>
<th>60 D</th>
<th>78 D</th>
<th>90 D</th>
</tr>
</thead>
<tbody>
<tr>
<td>Correction</td>
<td>0.88</td>
<td>1.11</td>
<td>1.33</td>
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**The Cup disc ratio:**
The cup/disc ratios in normal eyes are significantly larger horizontally than they are vertically. (In less than 7% of normal eyes, the horizontal cup/disc ratio is smaller than the vertical one).

This indicates that the quotient of the horizontal-to-vertical cup/disc ratios is usually higher than 1.0. It is important for the early diagnosis of glaucoma, in which, in the early to medium advanced stages, the vertical cup/disc diameter ratio increases faster than the horizontal one. This leads to decrease of the quotient of horizontal-to-vertical cup/disc ratios to values lower than 1.0. The cup/disc ratios are low in small optic nerve heads and they are high in large optic discs. An unusually high cup/disc ratio, therefore, can be physiologic in eyes with large optic nerve heads, while an average cup/disc ratio is uncommon in normal eyes with small optic discs. Normally a difference of 0.1 to 0.2 may be encountered between the two eyes. A difference of 0.3 indicates a probable damage.

**B. The retinal nerve fibre layer (RNFL):**
Glucomatous optic nerve atrophy is associated with an optic nerve fibre loss and thus decreased visibility of the RNFL. Four types of RNFL defects can be seen: (Figure-8)

1) Slitlike or groovelike defects which extend to disc margin. Usually no abnormalities of optic disc and visual field are present.
2) Wedge-shaped defects: where a localized defects of the RNFL are seen as wedge-shaped and not spindle-like defects, running towards or touching the optic disc border. If they are pronounced, they can have a broad basis at the temporal raphe of the fundus and its tip touching the optic disc border in less than 60° of the disc circumference and often a corresponding small deep visual field defect. These defects appear as well-outlined dark wedge-shaped areas in the bright striated pattern of surrounding healthy nerve fibre layer. Unlike slit-like changes, localized NFL defects rarely occur in normal eyes. (Figure-9).

In early glaucoma, bundle defects in the NFL may not affect the neuroretinal rim appearance because the damaged NFL is located in the deep retinal layers. Localized RNFL defects are not pathognomonic for glaucoma, since they occur also in other types of optic nerve atrophy, such as optic disc drusen, toxoplasmic chorioretinal scars, ischemic retinopathy with cotton-wool spots of the retina, Leber’s hereditary optic neuropathy, longstanding papilledema and optic neuritis due to multiple sclerosis.

3) Diffuse or generalized RNFL loss is the most common but the most difficult to detect. It may be noticed by evaluating whether the retinal vessels are clearly and sharply detectable. The retinal vessels are normally embedded in the RNFL. In eyes with a diffuse RNFL loss, the retinal vessels are covered only by the inner limiting membrane, resulting in a better visibility and a sharper image of the large retinal vessels. (Figure-10)

4) Combined localized damage with diffuse atrophy may be noticed with progression of glaucoma. RNFL evaluation can be performed with ophthalmoscopy, using bright light and the red free filter of the ophthalmoscope or the slit lamp. Fundus photography with red free light is quite helpful.

![Figure 8](image)

*Fig.(8)*: Retinal nerve fibre layer. (A) Normal, (B) Slit defect, (C) Wedge defect, (D) Diffuse atrophy.
When evaluating the RNFL the following points are to be considered:

1) If large blood vessels covered by bright reflexes near the disc margin, NFL is probably normal.

2) If secondary or tertiary vessels not covered by bright reflexes of the NFL, then diffuse (probably severe) RNFL damage is present.

3) Comparisons of superior-inferior areas in the same eye and similar areas in the fellow eye are very helpful.

Sharp demarcation lines and darker wedge-shaped swatches are evidence of localized RNFL abnormality. The apex of dark wedges should point toward the disc margin, typically near the inferior or superior poles.

Difficulties in evaluating the RNFL are encountered in presence of cataract, myopia and blonde fundus.
References:


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