Is the Optic Disc Cupping or Sinking in Glaucoma?

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Introduction

My presentation, 'Is the Optic Disc Cupping or Sinking in Glaucoma' is based on my personal affliction and observation of glaucoma patients. During my residency about 40 years ago, I was found to have a high intraocular pressure (IOP) of about 30 mmHg in each eye. I did not receive any treatment. I still have the same high IOP. Yet, I see 20/20, have no visual field defects, and my optic discs are still healthy.
It was very puzzling as to why I didn't develop glaucoma at a high IOP of 30 mmHg while others did at normal IOPs. This presentation will attempt to explain this puzzling question and how I arrived at the conclusion that the optic disc may not be cupping, but instead sinking in its entirety.

Background

What is intraocular pressure?

The eye contains a clear liquid known as aqueous humor which is produced by the ciliary body. After providing nutrition, the aqueous leaves the eyeball via the anterior chamber angle into the trabecular meshwork and rejoins the blood circulation. Since the aqueous is enclosed within the eyeball, it creates pressure which is known as intraocular pressure. Normal range is considered to be between 10-22 mmHg.

What is glaucoma?

There are various ways to classify glaucoma. For this presentation, glaucoma is classified into two types: painful and non-painful. The painful type is usually acute in onset in which there is a sudden rise of IOP to very high level of about 50 mmHg or more. Acute painful glaucoma is usually either due to a sudden blockage at the anterior chamber angle or in the pupillary area. In acute glaucoma, the painful eye is also inflamed and congested. If not treated urgently, it leads to blindness rapidly.

In the non-painful type of glaucoma, the eye is quiet and non-inflamed. The non-painful type of glaucoma is more common and was termed as simple glaucoma about 150 years ago. In simple glaucoma, there is a gradual loss of vision over many years.

In the past 40 years or so, another type of non-painful glaucoma has been recognized and termed as low or normal-tension glaucoma (NTG). In NTG, the intraocular pressure is consistently within the normal range, but has similar pathological changes in the optic disc and visual field defects as in the case of high-tension glaucoma (HTG) in which the IOP is above the normal range. It is believed that NTG is due to undue sensitivity of the optic disc to intraocular pressure therefore the optic disc atrophies even at a normal range of IOP. Some believe that NTG patients have a thin cornea therefore we are underestimating their true high IOP. Both HTG and NTG have open anterior chamber angle therefore they are also known as primary open angle glaucoma. This presentation is regarding these primary open-angle glaucomas.

What is the physiological and glaucomatous cup?

The physiological cup is the fibrous tissue base of Bergmeister's papilla which has remained after its atrophy in fetal life. The cup varies in size from 0.0 to 0.9 which is described as the cup-to-disc ratio. The term glaucomatous cupping implies when the physiological cup begins enlarging in glaucoma. After the invention of the ophthalmoscope in 1851 by Von Hemholtz, the ophthalmologists at the time were able to see the optic discs of simple glaucoma patients. They described these optic discs as 'cupped'. For the past 150 years, the term 'cupping' has become synonymous with glaucoma.

Nerve Fiber Arrangement

Retinal nerve fibers are the axons of the ganglion cells of the retina. There are about one million nerve fibers which converge on the optic disc. The arcuate fibers originate from the peripheral temporal retina and arch above and below the macular fibers to reach the optic disc. The arcuate fibers are of special interest because they are selectively destroyed in the earlier stages of glaucoma.

Visual Fields

In 1889, Jannik Bjerrum discovered isolated scotomas in the paracentral region on the perimetery in glaucoma. His pupil, Ronne, later found these isolated scotomas coalescing to form arcuate field defect ending at the horizontal raphe known as Ronne's nasal step. These arcuate field defects are produced by the selective destruction of the arcuate fibers in the initial stages of glaucoma. The central fields last (macular fibers) until the end stage of glaucoma.

The Puzzling Question

Why do some people develop glaucoma at a normal IOP such as 15 mmHg, while others not at a high IOP such as 30 mmHg?

Some physicians suggest that NTG patients have undue sensitive optic disc to IOP therefore the disc atrophies even at normal range IOP. Others believe that NTG patients have thin corneas therefore we are underestimating their true high IOP. Detailed medical history of glaucoma patients revealed that patients with HTG were usually in good health whereas, the patients with NTG had cardiovascular, circulatory and respiratory problems. Surprisingly, about 70% of the patients with NTG were long-term smokers. These findings suggested that NTG may be a systemic disease whereas, HTG, a disease of the eyeball itself. This findings also suggest that chronic glaucoma may be a multifactorial disease.

Now the question arises, If HTG is a disease of the eyeball itself whereas while NTG a systemic disease, then why are there similar glaucomatous changes in the optic discs and visual field defects of these two different types of glaucoma? I reasoned that there had to be a **common ground** somewhere in the course of the pathogenesis of HTG and NTG if glaucoma is indeed a multifactorial disease. I realized that the arcuate field defects, being the pathognomonic feature in both HTG and NTG, may provide a good lead in finding the common ground. Thus, any location or factor which could not possibly cause selective destruction of the arcuate fibers was ruled out in search for the common ground.

Hypothesis and Approach

The following questions pertain to the selective destruction of the arcuate fibers which are a pathognomonic feature of both HTG and NTG.

Can the arcuate fibers be selectively destroyed if 'cupping' of the optic disc is occurring?

Not likely: The cup is made of fibrous glial tissue and not elastic tissue. If the physiological cup is not elastic, why should it enlarge in response to raised IOP and then reverse in size when the IOP is lowered? If cupping is pressure induced, then why is there no cupping in acute glaucoma where the IOP reaches a very high level while there is cupping present in NTG where the IOP is in normal range? Even if the physiological cup is enlarging, how is it possible that glaucomatous cupping involving 360 degrees could selectively destroy the arcuate nerve fibers and not encompass the rest of the nerve fibers? If this is not possible, then the phenomena of cupping may not be occurring in glaucoma.

Can the arcuate fibers in the optic disc be selectively destroyed by any cause?

Not likely: The optic disc is composed of about one million nerve fibers which are densely packed in the 1.5 mm size disc. How is it possible that raised IOP, or any other pathology, could selectively target the arcuate fibers and not the rest in such a highly dense optic disc? If this is not possible, then the optic disc may not be the primary site of injury.

Can the arcuate fibers of the retina be selectively destroyed by any cause?

Not Likely: How is it possible that raised IOP or any other pathology could selectively destroy the retinal nerve fibers among the rest of the 360 degrees of nerve fibers? Regarding genetically controlled apoptosis, how is it possible that when apoptosis is activated, that it will initiate selectively only with those ganglion cells which serve the arcuate fibers and not randomly as it occurs in the rest of the body? How can we explain the selective destruction of the arcuate fibers occurring in chronic traumatic and secondary glaucoma which are not genetically controlled? If this is not possible, then the retina may not be the primary site of injury.

Can the border tissue be the primary site of injury in glaucoma?

If optic disc or the retina can't be the primary site of injury, then we are left with the border tissue. The circular Border Tissue of Elschnig separates the scleral edge from the optic disc and also acts as an 'O' ring sealant which keeps the optic disc firmly in place. The optic disc has a dual source of blood supply, central retinal artery (CRA) and short posterior ciliary arteries (SPCA's). However, the circular border tissue is exclusively supplied by the SPCA's. Unfortunately, the central retinal artery does not take part in its blood supply. The circulation of the SPCA's is a low pressure system due to multiple branches of short posterior ciliary arteries.

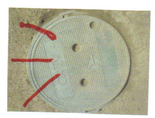
The intraocular pressure and arterial pressure are opposing forces. Normally, IOP equates the arterial pressure of the border tissue for its survival. Circulation and oxygenation of the border tissue would be decreased either by increased IOP or due to poor systemic circulatory and respiratory problems and smoking. In both instances, there would be a chronic lack of perfusion of the border tissue resulting in its atrophy. **Therefore, the border tissue may be the common ground in the pathogenesis of HTG and NTG.**

What happens after the border tissue atrophies?

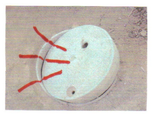
Due to weakening of the border tissue, the optic disc would begin sinking in the scleral foramen. An analogy would be a road made of nerve fibers and the manhole cover as the optic disc. If the manhole cover begins sinking due to its weak attachments, then the road nerve fibers would be stretched and severed at the edge due to a break in surface continuity. I believe that this likewise process may be occurring to the optic disc in glaucoma (Diagram A).

Can the arcuate fibers be selectively destroyed if the optic disc is sinking?

Likely: Our genes have already separated the arcuate fibers from the rest of the nerve fibers. Secondly, the optic disc has an oblique entry, therefore, its temporal part is closer to the scleral edge. As the disc sinks, the temporal fibers, which include superior and inferior arcuate and the centrally located macular fibers, would be stretched and severed at the edge. Since the arcuate fibers are much less in number as compared to the macular fibers, the arcuate fibers would be depleted earlier giving rise to double arcuate field defects.
The macular fibers being abundant, will deplete last, therefore, the central vision will be retained until the end stage of glaucoma (Diagram B).

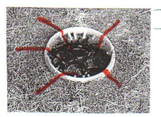


Normal: Flush with road

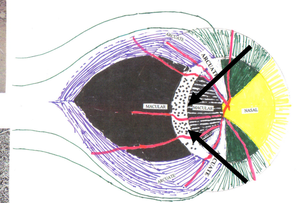


Early stage of sinking

Middle Stage



Final Stage



Arcuate fibers being cut at the scleral edge due to sinking of the temporal part of the optic disc.

Diagram A: Sinking Manhole Cover

Diagram B

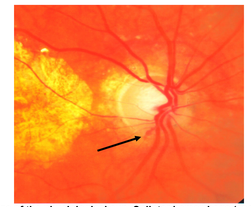
What happens as the sinking of the disc progresses?

The retinal nerve fibers anchor the optic disc in place as roots anchor a tree. As the nerve fibers are being severed and depleted, the optic disc becomes more loose and sinks further resulting in severing of the additional nerve fibers. This would create a self propagating cascade of loosening and sinking which would continue until all the nerve fibers are severed at the edge and the disc is totally perished. The area which previously housed the optic disc is replaced with an empty crater with the larger blood vessels hanging on the edge.

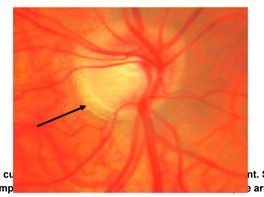
Conclusion

The optic disc may not be cupping but instead sinking in its entirety. The physiological cup may not be enlarging, but instead breaking up due to merger of the cup pallor with the pallor produced by the destruction of the nerve fibers and its vasculature. Normal tension glaucoma may be a systemic disease and glaucoma itself a multifactorial disease. Furthermore, the more risk factors present, the higher the likelihood and severity of the development of glaucoma akin to ischemic heart disease. The nerve fibers are being severed and depleted. Finally, at the end stage, there is no optic disc. This is what the histology of the end stage glaucomatous disc reveals.

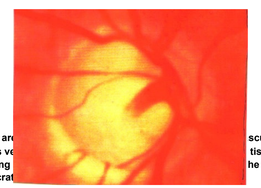
Figures



Early Stage. No change in size of the physiological cup. Splinter hemorrhage (see arrow) present at the 7 o'clock position. Presence of temporal pallor and prominent scleral edge due to thinning of the nerve fiber layer. Sloping and kinking of the vessels at edge (right eye).



Middle Stage. Physiological cu nt. Sloping of temporal blood vessels. Baldness at the temp arrow). Beginning of nasal shifting of the vessels as a result of loss of anchorage due to depleted temporal fibers. (right eye)



Final Stage. The optic disc ar sculature. The kinking of the blood vessels becomes ve tissue. Entire scleral foramen is visible. Marked nasal shifting he temporal fibers. Optic disc area is replaced with an empty crat

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