IDIOPATHIC MACULAR HOLE
DIAGNOSIS AND MANAGEMENT

Essay

Submitted for Partial Fulfillment of Master Degree in
Ophthalmology

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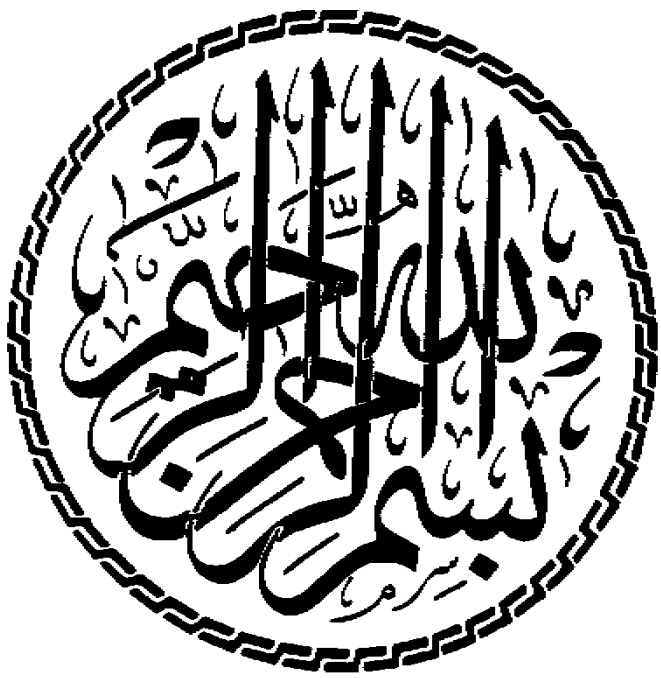
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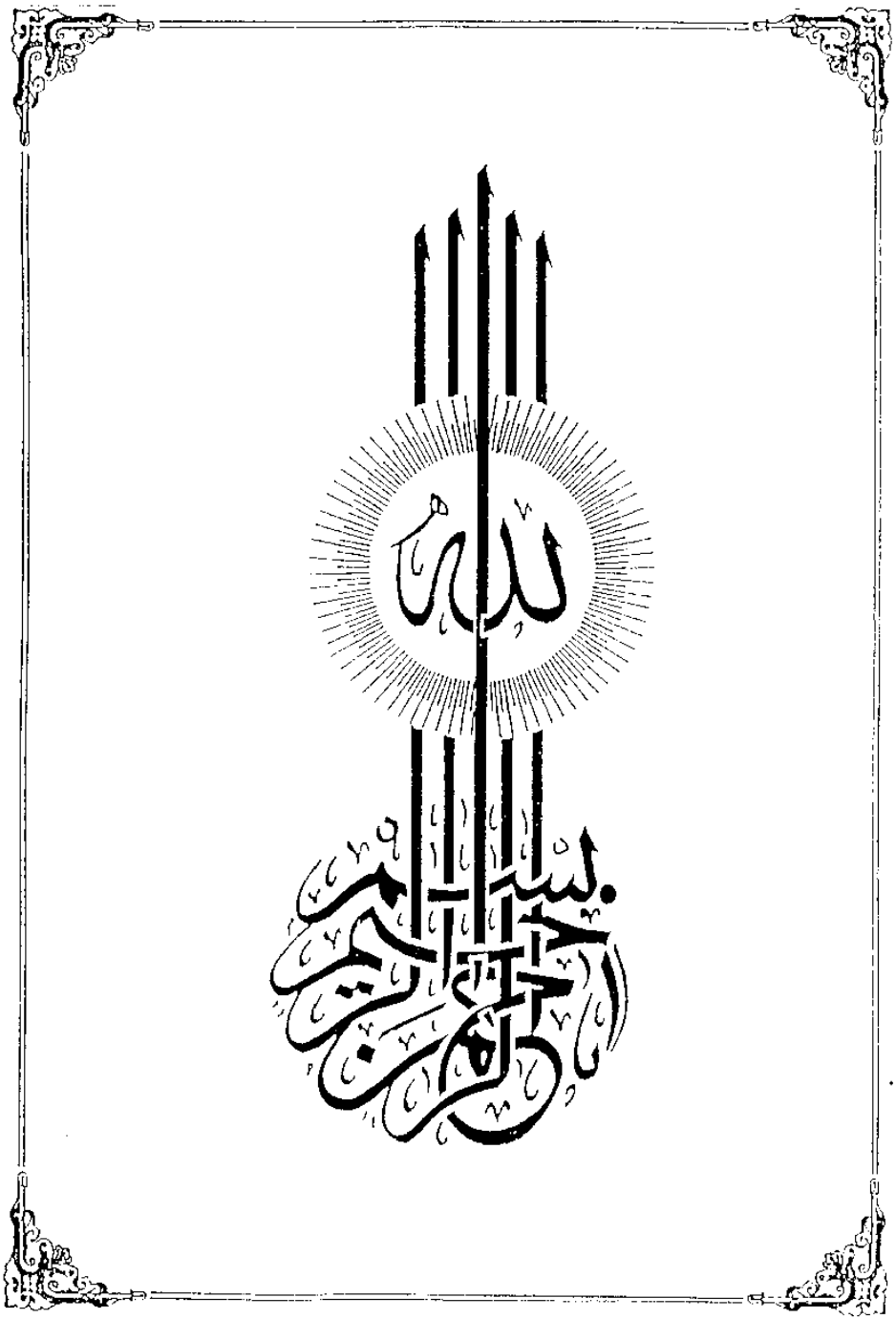

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Sherif Emerah

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To my Parents

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NORMAL MACULA

Anatomy of the Normal Macula

Definition

Anatomically the macula is defined as that portion of the posterior retina that contain xanthophyllic pigment and two or more layers of ganglion cells. It measures about 5.5 mm in diameter and is centered approximately 4 mm temporal to and 0.8 mm inferior to the center of the optic disc.

Anatomic Subdivisions

The macular area can be further subdivided into several zones. The fovea (fovea centralis) is a depression in the inner retinal surface in the center of the macula. It measures about 1.5 mm or one disc diameter in size. The central floor of the fovea is called the foveola. It measures about 0.35 mm in diameter. It lies within capillary-free zone, which measures about 0.5 mm in diameter. A small depression in the center of the foveola is called the umbo. A 0.5 mm wide ring zone where the ganglion cell, innernuclear layer, and outer plexiform layer of Henle are the thickest is called the parafoveal area. This zone is in turn surrounded by a 1.5 mm zone referred to as the perifoveal area *(Gass, 1987) (See figure 1).*

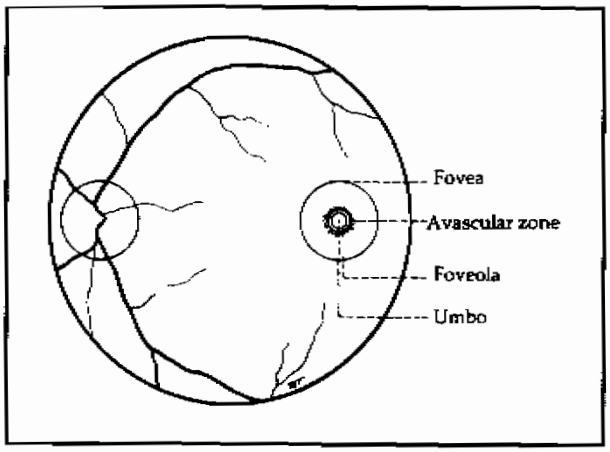

Figure 1

Anatomy of the Fovea *(Kanski, 1989)*

Gross Anatomy

Xanthophyllic pigments are yellow pigments apparent in the center of the macula, its concentration are maximal in the area corresponding with the foveola. If a fresh human retina in the central macular area is viewed in cross section, the concentration of these pigments appears to be maximal in the outer nuclear and outer plexiform layers. It is also present within the inner plexiform layer inside the foveal area. Its precise location histologically has never been determined. It has an absorption spectrum consistent with that of lutein, an alpha-hydroxy carotene, and has been extracted biochemically from the human retina as xanthophyll. The relatively dark area in the foveal region is probably caused primarily by the increased pigment content of the RPE cells. Some of the relative darkness of the central macular area remains, however, because of the greater concentration of the choroidal melanocytes in this area.

The short posterior ciliary arteries are concentrated in the macular area, particularly along the temporal margin of the fovea and the

peripapillary area. They branch frequently and course outward toward the periphery. Several short posterior ciliary arteries enter nasal to the optic disc. The temporal long posterior ciliary artery and ciliary nerve enter about one and one-half disc diameters temporal to the center of the fovea. Melanocytes are concentrated along either side of these structures *(Gass, 1987)*.

Clinical Appearance

The anatomic subdivisions of the macula are ill defined. The center of the macula is usually evident as poorly defined zone of slightly greater pigmentation that is maximum in the foveolar area. The foveal reflex, which is present in most normal eyes, appears to lie just in front of the center of the foveola and therefore overlies the anatomic umbo *(See figure 2)*. There are no ophthalmoscopic landmarks to indicate the margins of either the 0.35 mm diameter foveola or the 1.5 mm diameter fovea. In younger patient a halo like reflex may correspond with the foveal margin. The foveal depression can be visualized with the narrow slit-lamp beam *(Gass, 1987)*.

Figure 2

Normal Foveal Light Reflex *(Kanski, 1989)*

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Figure 2

Normal Foveal Light Reflex *(Kanski, 1989)*

The macular region is supplied by the superior temporal and inferior temporal branches of the central retinal artery. It is found in 20% of patients that variable portion of the papillomacular area was supplied by one or more cilioretinal arteries derived from the ciliary circulation. Occasionally a large cilioretinal artery may supply the entire macula *(Justice and Lehmann, 1976).*

The retinal pigment epithelium (RPE) is more highly pigmented in the central macular area than elsewhere. In white persons it imparts an orange or orange-red color to the fundus in most black persons it imparts a brown color *(Gass, 1987).*

Histology

Photoreceptors cells, both rods and cones, are specialized, elongated cells whose nuclei comprise the outer nuclear layer of the retina.

Both rods and cones have specialized light gathering ends. The outer segment, which consist of stacks of flat lipid-bilayer discs. The inner segment, which connect the discs to the cell body, are filled with mitochondria and other organelles of synthesis and transport.

In the foveola the only photoreceptors are cones, although they are long, slender, rod-like, and densely packed. Only the cones and some Müller cells are present in the foveola, the remaining layers of the sensory retina having been pushed aside *(Bok, 1985).*

The photoreceptors form synapses with the cells of the inner nuclear layer. The outer plexiform layer lies between the outer and the inner nuclear layers, and is the site where the synapses take place. The cell processes extending from the cones in the foveola are obliquely oriented forming Henle's fiber layer *(See figure 3)*. Fluid can more easily collect in this region of the outer plexiform layer because the obliquely oriented fibers are more readily displaced than the vertically oriented fibers in the peripheral retina *(Hogan, Alvarado, and Weddell, 1971)*.

Figure 3

Cross-Section of Fovea *(Kanski, 1989)*

The inner nuclear layer is comprised of the cell bodies of horizontal, bipolar, and amacrine cells. These cells in turn form a synapse with the ganglion cells in the inner plexiform layer.

The ganglion cell layer is the innermost layer of cell nuclei. These cells are relatively large, with abundant cytoplasm. The anatomic macula is defined as that area where the ganglion cell layer is two or more cells in thickness, and more centrally it can be up to seven or eight cells thick. The ganglion cells send a single axon through the nerve fiber layer to the optic nerve. The next synapse is located in the lateral geniculate body *(Hogan, Alvarado, and Weddell, 1971)*.

The large arterioles and venules of the retinal circulation travel in the nerve fiber layer and ganglion cell layer. Close to the foveal vascular zone the capillaries form a single layer, but elsewhere the capillaries are present in two or more layers and extend outward into the inner nuclear layer.

The inner limiting membrane is formed in part from a true basement membrane secreted by the Müller cells. These cells are elongated cells that extend nearly the full retinal thickness, from the photoreceptors nuclei on inward *(Kincaid, 1993)*.

The microvilli processes of the retinal pigment epithelium surround the photoreceptors outer segments which have been shown to be shed and regenerated constantly throughout life at a rate dependent on incident light. The shed material is engulfed and digested by the retinal pigment epitheluim. There are no structural junctions between the retinal pigment epithelial cells and the photoreceptors. The retinal pigment epithelium constantly pumps fluid from the subretinal space to maintain photoreceptors apposition. This active transport accounts for about 70% of the total forces responsible for retinal apposition *(Kincaid, 1993)*.

Macular Function Tests

Visual Acuity

Visual acuity measurement is the most common test of foveal function. The eye must first be refracted to best correction for acuity to be a significant measure of foveolar performance. Acuity is measured by the visual resolution of a letter, symbol, or a pattern. The smaller the visual angle subtended by the parts of the test letter correctly seen by the observer, the better the resolution of his visual system. A second factor involved in acuity measurement is contrast. Visual resolution of single figure generally suffers when contrast is markedly diminished *(Wolkstein and Carr, 1979)*.

The Amsler Grid

This test evaluates the 10° of visual field surrounding fixation. It is primarily used for screening of macular disease. The chart consists of a 10 cm square divided into smaller 5mm squares *(See figure 4)*. When it is viewed at one-third of a metre, each small square subtends an angle of 1°. The test is performed as follows:

a) The patient should wear reading glasses and cover one eye.

b) The patients is asked to look directly at the center dot with the uncovered eye and report any distortion, wavy lines, blurred areas or blank spot anywhere on the grid *(Kanski, 1994)*.