

Ophthalmologic Manifestations In Rheumatic Diseases

Essay

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List of Abbreviations

A-AION	Arteritic Anterior Ischemic Optic Neuropathy
ACAID	Anterior Chamber – Associated Immune Deviation
AIDS	Acquired Immune Deficiency Syndrome
AION	Anterior Ischemic Optic Neuropathy
MSH	Melanocyte Stimulating Hormone
ANCA	Antineutrophilic Cytoplasmic Antibody
APCs	Antigen Presenting Cells
ARN	Acute Retinal Necrosis
CD	Cluster of Differentiation
CTL	Cytotoxic T-Cells
DS	Diffuse Scleroderma
DTH	Delayed Type Hypersensitivity
EBV	Epstein Barr Virus
EMC	Essential Cryoglobulinemic Vasculitis
EUA	Experimental Autoimmune Uveitis
GCA	Giant Cell Arteritis
GM-CSF	Granulocyte- Macrophage Colony Stimulating Factor
HLA	Human leukocyte Antigen
HTLV	Human T- lymphotropic Virus
IBD	Irritable Bowel Disease
IL	Interleukin
INF	Interferon
JAS	Juvenile Ankylosing Spondylitis
JIA	Juvenile Idiopathic Arthritis
KCS	Keratoconjunctivitis Sicca
KD	Kawasaki Disease
MHC	Major Histocompatibility Complex
MPA	Microscopic Polyangitis
NA-AION	Non - Arteritic Anterior Ischemic Optic Neuropathy
NK cells	Natural killer Cells
NOMID	Neonatal Onset Multisystem Inflammatory Disease
ODE	Optic Disc Edema
ONH	Optic Nerve Head

List of Abbreviations

PAN	Polyarteritis Nodosa
PCA	Posterior ciliary artery
PCR	Polymerase Chain Reaction
PD	Programmed Death
PDL	Programmed Death Ligands
PION	Posterior Ischemic Optic Neuropathy
PSC	Posterior Subcapsular Cataracts
PUK	Peripheral Ulcerative Keratitis
RA	Rheumatoid Arthritis
RPE cells	Retinal Pigment Epithelial Cells
SLE	Systemic Lupus Erythematosus
SpAs	Spondyloarthropathies
SPCA	Short Posterior Ciliary Artery
SS	Sjogren syndrome
SS-ATD	Sjogren syndromes aqueous tear deficiency
SSC	Sicca syndrome
TAK	Takayasu arteritis
TGFb-2	Transforming growth factor – beta2
TH	T-helper cells immune response
TNF	Tumor necrosis factor
TRAIL	TNF related apoptosis – inducing ligands
Treg cells	Regulatory T cells
TSP	Thrombospondin
WG	Wegner granulomatosis

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Introduction

Rheumatic diseases represent a group of autoimmune conditions which primarily affect the musculo-skeletal system but can also involve other internal organs such as the ocular, auditory and respiratory systems and therefore exhibit a wide spectrum of clinical manifestations **(Rachel, 2010)**.

The eye is an especially sensitive barometer for the onset or flare up of autoimmune phenomena in many of the rheumatic diseases **(Fayad and Pamela, 2001)**.

Ophthalmic manifestations may occur during the course of rheumatic diseases like rheumatoid arthritis, Systemic lupus erythematosus, juvenile rheumatoid arthritis, Sjogren syndrome, Scleroderma, Behcet's s., Vasculitis (especially Wegner's and Giant cell arteritis) and antiphospholipid antibody syndrome **(Carl ,2010)**.

Ocular complications occur in approximately 25% of patients with rheumatoid arthritis (RA) like dry eye syndrome (keratoconjunctivits sicca, KCS), episcleritis, scleritis, and corneal ulceration **(Russell and Suzanne, 2010)**.

Systemic lupus erythematosus (SLE) can affect the eye, optic nerve, and ocular adnexa. The most

common ocular findings include dry eye and retinal vascular changes (**John and Prabakar, 2008**).

In Sjogren's syndrome, Keratoconjunctivitis sicca (dry eyes) is the hallmark sign. Sicca syndrome is defined as KCS and xerostomia without any extraglandular disease (**Petra et al., 2010**).

Behcet's disease may presents with findings including, cataract, posterior synechiae and posterior capsular opacification secondary to cataract surgery, vitreous condensation, optic atrophy, cystoid macular edema, retinal periphlebitis, neovascularization, and phthisis bulbi (**Mehmet and Nilufer, 2009**).

In vasculitis known as Antineutrophilic cytoplasmic antibody (ANCA) positive which is a term that is favored to designate the disease otherwise known as Wegener's granulomatosis we see findings like scleritis, peripheral keratitis, orbital pseudotumor, retinal vasculitis , and neuro-ophthalmic lesions (**Angela et al.,2009**).

In scleroderma ophthalmic findings include, eyelid scleroderma, conjunctival telangiectasia, and dry eye (**Baroni et al., 2006**).

In ankylosing spondylitis ophthalmic complications are presented in the form of acute anterior uveitis (**Mark, 2007**).

Also Ophthalmic manifestations may occurs as a side effect of medications used for treatment of rheumatic diseases , every part of the eye and all ocular functions could be affected adversely. Like nonsteroidal anti-inflammatory, anti-malarial drugs which may leads to retinopathy, Corticosteroids that may induce cataract or glaucoma, and others may induce side effects (**Hafstrom et al., 2009**).

Because many of these ocular complications may result in loss of vision, evaluation by an ophthalmologist is needed and his coordination with the rheumatologist can play a major role in detecting and managing the eye involvement in his patients to save this important sense. Understanding the varied manifestations of eye disease will permit the rheumatologist to better evaluate the activity of the rheumatic disease (**Fayad and Pamela, 2001**).

Aim of the work

To highlight the ophthalmologic disorders of rheumatic diseases as regard early diagnosis and management to prevent complications.

Human Eye

Eyes are organs that detect light and convert it to electro-chemical impulses in neurons. The simplest photoreceptors in conscious vision connect light to movement. In higher organisms the eye is a complex optical system which collects light from the surrounding environment, regulates its intensity through a diaphragm, focuses it through an adjustable assembly of lenses to form an image, converts this image into a set of electrical signals, and transmits these signals to the brain through complex neural pathways that connect the eye via the optic nerve to the visual cortex and other areas of the brain. Eyes with resolving power have come in ten fundamentally different forms, and 96% of animal species possess a complex optical system. Image-resolving eyes are present in molluscs, chordates and arthropods (Ang, 2008).

1) Surface Anatomy of Human Eye (Moore et al, 2007).

In health, only a small part of the eye is exposed anteriorly (fig.1). Seven-eighths of the cornea and a small area of the nasal and larger area of the temporal sclera are visible when looking straight ahead. Viewed from the side, a considerable portion of the globe (17 mm on average) is anterior to the lateral orbital margin, but this is very variable and may be much less with age.

Outside the floor of the orbit are the maxillary sinus and the infra-orbital nerve. The two lateral walls are at right angles to each other. The optic nerve and globe are centred on the orbital axis which is 22.53 lateral to the sagittal axis. Looking straight ahead (the primary position) the optic axis is at 22.53 to the orbital axis.

The projecting brow, nasal bones and zygomatic arch provide protection to the globe. The eye measures 23 mm on average from front to back and slightly less equatorially. The intra-orbital optic nerve measures about 25 mm. The distance between the back of the eye and the apex of the orbit is slightly less (the optic nerve takes a sinuous course to allow for eye movement). The distance between the centers of the orbits varies considerably but is around about 65 mm on average.

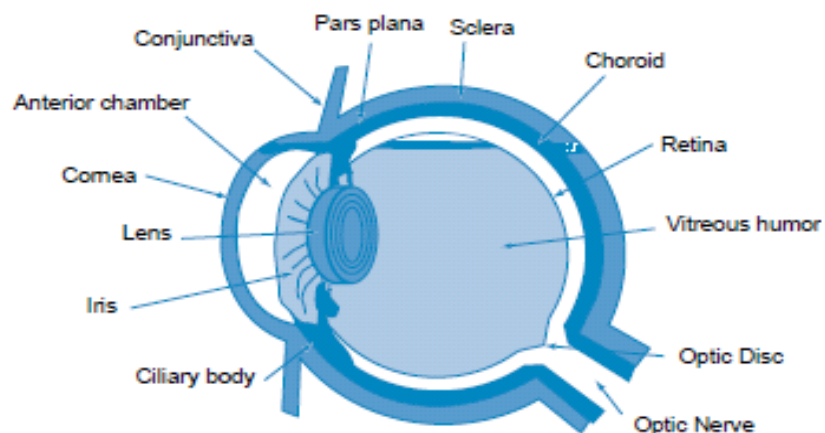


Fig. 1: The globe (Santiago and Emilio 2006).

2) Structure of Eye:

The Protective Structures of Eye

• The Orbit:

The two orbits, sometimes referred to as “sockets,” that protect the human eyes are situated at the front of the skull; each orbit is roughly pyramidal with a wider opening to the front narrowing to a small opening at the rear where the optic nerve exits to connect through the visual pathways and the brain (fig.2). The orbits are angled outward approximately 23° with respect to the midline of the skull. The human eye itself is approximately 24 mm in diameter and occupies about 25% of the volume of the orbit, allowing for the extra ocular muscles, blood vessels, nerves, orbital fat and connective tissue that surround and support the eye (Ang, 2008).

Behind the apex of the orbit are the cavernous sinus and the important structures of the midbrain. The thin medial walls of the orbit are parallel to each other. Between the medial walls are sinuses, the nasal cavity and the nasolacrimal system. Outside the lateral wall is the temporal fossa, which houses the temporalis muscle. Outside the roof of the orbit is the frontal lobe of the brain posteriorly and the frontal sinus anteriorly (Delcourt et al., 2000).

It Surrounds and supports most of the human eye, while the cornea and part of the anterior globe extend somewhat beyond the orbital rims. These structures are protected by the globe itself is predominately formed of and protected by the sclera that extends from the edges of the clear cornea at the front of the eye (the “limbus”) to the optic nerve at the back of the eye (**Congdon and Friedman, 2003**).

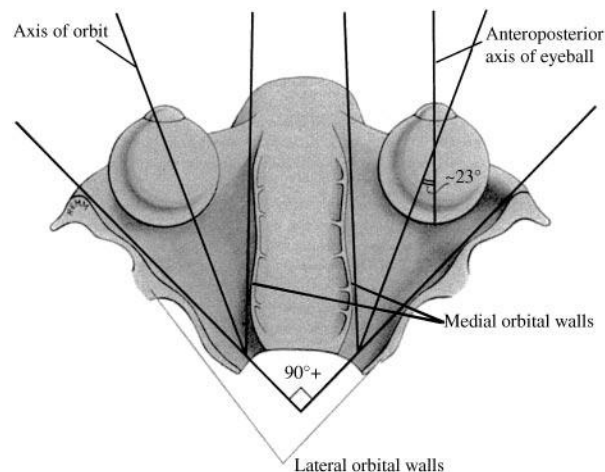


Fig.2: The general arrangement of the orbit (**Kersten, 2000**).

• The Conjunctiva:

The conjunctiva is the thin translucent mucous membrane of the eye, which is part of the maintenance visual system of the ocular surface, serves as the junction between the eyeball and the eyelids. Its most superficial epithelial layer is totally covered by the tear film. It is continuous with the nasal mucosa through the