Uveitis masquerade syndromes

Essay protocol submitted for partial fulfillment of the Master Degree in Ophthalmology

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List of Contents

List of Abbreviations

ACE	**Angiotensin-converting enzyme**
AIDS	Acquired immunodeficiency syndrome
ANA	Antinuclear antibody
APMPPE	Acute posterior multifocal placoid pigment epitheliopathy
ARN	Acute retinal necrosis
CAT	Computed axial tomography
CC	Cubic centimeter
CHOP	Cyclophosphamide, Hydroxiurea, Oncovine, Prednisolone .
CME	Cystoid macular edema
CMV	Cytomegalovirus
CNS	Central nervous system
COMS	Collaborative Ocular Melanoma Study Group
CRVO	Central retinal vein occlusion
CSF	Cerebrospinal fluid
DHA	Docosohexanoic acid
ELIZA	Enzyme linked immunosorbant assay
ERG	Electroretinogram
FA	Flourescein angiography
FDA	Food and Drug Administration
FTA	Fluorescent Treponemal antibody
G	Gram
GI	Gastrointestinal
HLA	Human leukocyte antigen
HSV	Herpes simplex virus
HZV	Herpes zoster virus
ICGA	Indocyanine green angiography
INF	interferon

IL	Interleukin
IOP	Intraocular pressure
JIA	Juvenil idiopathic arthritis
JXG	Juvenile xanthogranuloma
Kg	Kilogram
KPs	keratic precipitates
MEWDS	Multiple evanescent white-dote syndrome
Mg	Milligram
MRI	Magnetic resonance imaging
MRA	Magnetic Resonance Angiography
NF-AT	Nuclear factor of activated T lymphocytes
NSAIDS	Nonsteroidal anti-inflammatory drugs
OCT	Optical coherence tomography
OIS	Ocular ischemic syndrome
PCNSL	Primary central nervous system lymphoma
PCR	Polymerase chain reaction
PERG	pattern ERG
PET	Positron emission tomography
PO	Per oral
RP	Retinitis pigmentosa
RPE	Retinal pigment epithelium
RPR	Rapid plasma reagent
SUN	Standardization of Uveitis Nomenclature
TINU	Tubulointerstitial nephritis and uveitis
VA	Visual acuity
VKH	Vogt-Koyanagi-Harada Syndrome
VZV	Varicella-zoster virus

List of Tables

List of Figures

INTRODUCTION:

Uveitis is defined as inflammation of the uveal tract, the vascular coat of the eye which is composed of the iris, ciliary body and choroid. Inflammation of these structures is frequently accompanied by involvement of the surrounding ocular tissues, including the cornea, sclera, vitreous, retina and optic nerve **(Foster & Vitale 2002).**

So the term uveitis is used clinically to describe a heterogeneous group of diseases characterized by inflammation of intraocular structures **(Suhler et al*, 2005*, Kanski J, 2007).**

The term *masquerade syndrome* was first described as a chronic case of conjunctivitis masquerading conjunctival carcinoma **(Theodore et al, 1967)**

Now it is classically used in ophthalmology to describe those conditions that include as part of their manifestation the presence of intraocular cells, but not due to immune-mediated uveitis entities **(Rothva et al, 2001 Read et al, 2002).**

In fact, Ocular masquerade syndromes are the most commonly used term to describe diseases simulating a chronic uveitis **(Nussenblatt et al, 2004).**

So they are a group of diseases that occur with ocular inflammation and are misdiagnosed as a chronic uveitis **(Nussenblatt et al, 2004).**

Also the term masquerade syndromes refer to ophthalmic disorders that are not primarily inflammatory in nature but may clinically present as either anterior or posterior uveitis.

These entities may be mistaken for, or masquerade as, primary uveitis. Extensive evaluation is often initiated as patients can manifest with atypical features such as recurrent episodes of uveitis, or uveitis unresponsive to standard

therapeutic measures. **(Vrabec et al, 2007).**

The causes of Ocular masquerade syndrome can be divided into neoplastic conditions and non-neoplastic conditions **(Rothva et al, 2001).**

The commonly encountered neoplastic masquerade syndromes include:

- Primary CNS Lymphoma (PCNSL).
- Secondary to Systemic Lymphoma.
- secondary to systemic leukemia
- Uveal Lymphoid Proliferations
- Uveal Melanoma
- Retinoblastoma
- Juvenile Xanthogranuloma
- Metastatic Tumors **(Read et al, 2002).**

The commonly encountered non-neoplastic masquerade syndromes include:

- Ocular ischemic syndrome.
- Chronic peripheral rhegmatogenous retinal detachment.
- Retinitis pigmintosa **(Ramana et al, 2008).**

AIM OF THE WORK

This essay aims to review the most common masquerade syndromes that can mimic primary uveitis.

Clinical approach to uveitis

Uveitis encompasses a myriad of conditions, all of which are characterized by inflammation of the uveal tract (iris, ciliary body, choroid), either directly or indirectly. The ophthalmologist's goal in treating these potentially blinding conditions is to eliminate the inflammatory reaction within the eye while minimizing the potential risks of therapy to the patient. This is best achieved once an accurate diagnosis has been obtained. To do this most efficiently, an extensive history and ophthalmologic examination are required (**Forster, 2008).**

After the physician has used the information obtained from the history and physical examination to determine the anatomical classification of uveitis, he or she can use several associated factors to further subcategorize, which leads in turn to choosing the laboratory studies. Laboratory studies help to determine the etiology of the intraocular inflammation, which leads to the selection and administration of theraputic options **(Ramana et al, 2007).**

EPIDEMIOLOGY AND PATHOGENESIS:

While numerous methods of classifying uveitis have been used in the past, the classification scheme recommended by the International Uveitis Study Group and the Standardization of Uveitis Nomenclature (SUN) Working Group is based on anatomic location (Table 2-1) **(SUN ,2005).**

In addition, descriptors of uveitis are used to further define the type of inflammation the patient exhibits. These include *onset* (sudden vs. insidious); *duration* (limited - less than 3 months' duration, persistent - greater than 3 months' duration); and *course* (acute, recurrent, or chronic). Additional features such as laterality (unilateral vs. bilateral) and granulomatous vs.

nongranulomatous appearance may also be helpful in determining an etiology for a patient's uveitis (**Forster, 2008).**

TABLE 2-1: THE CLASSIFICATION OF UVEITIS **(Bloch-Michel and Nussenblatt, 1987).**

Type	Primary Site of Inflammation	Includes
Anterior uveitis	Anterior chamber	Iritis-Iridocyclitis-Anterior cyclitis
Intermediate uveitis	Vitreous	Pars planitis-Posterior cyclitis-Hyalitis
Posterior uveitis	Retina or choroid	Focal, multifocal, or diffuse choroiditis-Chorioretinitis-Retinochoroiditis-Retinitis-Neuroretinitis
Panuveitis	Anterior chamber, vitreous, and retina or choroid	

Clinical picture of uveitis

Anterior uveitis:

Symptoms:

Acute anterior uveitis

Usually has a sudden, symptomatic onset and persists up to three months. If the inflammation recurs following the initial attack, it is referred to as recurrent acute **(Kanski, 2007)**.

Pain, redness, photophobia, consensual photophobia (pain in the affected eye when a light is shone in the fellow eye),

excessive tearing and decreased vision **(Justis and Chirag, 2008).**

Chronic anterior uveitis

It persists for longer than three months. The onset is frequently insidious and may be asymptomatic, although acute or subacute exacerbations may occur **(Kanski, 2007)**.

Decreased vision from vitreous debris, cystoid macular edema (CME), or cataract. May have periods of exacerbations and remissions with few acute symptoms [e.g. juvenile idiopathic (rheumatoid) arthritis] **(Justis and Chirag, 2008).**

Signs:

The conjunctiva classically shows perilimbal injection (known as ciliary flush). The cornea may have keratic precipitates, which are clusters of epithelioid cells, lymphocytes and WBCs collected on the endothelium. The type of keratic precipitate can provide a clue to the classification of anterior uveitis. Mutton-fat keratic precipitates are characteristic of granulomatous uveitis **(Fig. 2. 1).** Diffuse stellate keratic precipitates classically are seen in Fuchs heterochromic iridocyclitis. Interstitial keratitis commonly is seen in patients with syphilis and herpetic disease **(Smith and Nozik, 2003)**.

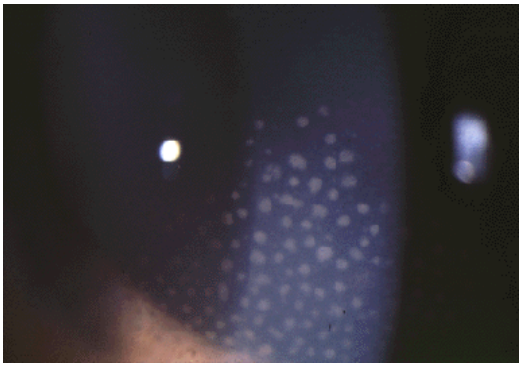

Fig.2.1: Keratic precipitates in anterior uveitis (mutton fat): KPs granulomatous in appearance, as would be expected with entities such as sarcoidosis, Vogt-Koyanagi-Harada syndrome, and sympathetic ophthalmia **(Forster, 2008).**