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Ocular Diagnostic clues in Retinal Vasculitis Uveitis course Antalya 2013

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### In this talk...

- Vasculitis affecting retinal arteries
- Vasculitis affecting retinal capillaries
  - Vasculitis affecting retinal veins

# Vasculitis – pathological definition

- Inflammatory (leucocyte mediated) destruction of blood vessel wall
- Arteritis, capillaritis, venulitis
- Localised or systemic (organ predilection)
- Neutrophil/lymphocytes/monocytes adhesion – activation transmigration necrosis (leucocytoclasis) or apoptosis

# Retinal vasculitis – clinical definition

Inflammation of retinal blood vessels

# The Standardization of Uveitis Nomenclature (SUN) Working Group

- Retinal Vasculitis is a descriptive term for situations where evidence of ocular inflammation and 'retinal vascular changes'
- However achieving consensus on which retinal vascular changes constitute retinal vasculitis was more problematic!

e.g.peripheral vascular sheathing vs leakage or occlusion on fluorescein angiography

Am J Ophthalmol 2005 140:509 - 516

Retinal vasculitis- suggestion based on pathology\*

• Classify retinal vasculitis based on involved vessels

arteries/arterioles

veins

post capillary venules/capillaries

- Associated with systemic disease
- Localised to retina

\*Narsing Rao Ettal Workshop 2008

### The spectrum of vasculitis Chapel Hill Consensus Conference 1994



Retinal arterial involvement without inflammation – Systemic vasculitides





### Choroidal infarcts in giant cell arteritis: Central





## Retinopathy of SLE











Central retinal artery occlusion in Lupus with APS



Peripheral choroidal infarcts in APS

In general systemic vasculitides do not cause uveitis Main presentation is arterial occlusive disease Retinal arterial involvement with inflammation – Viral/protozoal infections

### Acute retinal necrosis

- Is a clinical syndrome caused by the DNA viruses with retinal ARTERITIS and periphlebitis
- Herpes Simplex
- Varicella Zoster
- Cytomegalovirus
- Viruses par excellence preferentially affect ARTERIOLES





#### Varicella Zoster



CMV in chronically immunosuppressed for lung disease



#### **Courtesy NP Jones**



# Kyrieleis phenomenom affecting retinal arteries

### Acquired toxoplasmosis in immunodeficient



# Retinal Venous involvement with inflammation

## **Retinal VEINS**

- Can occlude major branches, smaller branches
- Can leak in a diffuse or focal manner
- When retinal veins are affected the main differential diagnoses are:
  - sarcoidosis, tuberculosis, multiple sclerosis,
    - Behcet's disease
      - idiopathic disease intermediate, ischaemic

### Sarcoidosis

- Periphlebitis may be mild or so severe that it may cause vascular occlusion classically appears like fluffy cuffing of retinal veins
- Involvement of post capillary venules may also occur





Sarcoidosis showing typical 'rat bite' periphlebitis – focal cuffing of veins with focal leakage







56 year old lady presents with mild panuveitis – she dies from myocardial infarction









### Tuberculosis in young African girl



# Eales disease due to TB







Pattern of uveitis in 30 patients with definite M.S. and symptomatic eyes

Anterior uveitis	74%
granulomatous	58%
Vitritis	74%
Retinal venous sheathing	84%
Retinal ischaemia	20%
Vitreous haemorrhage	20%
Phthisis	10%
RPE, choroid disease	0%







### Retinal vasculitis in Behcet's disease

- Major retinal vein occlusion
- Minor retinal vein occlusion
- Florid periphlebitis is NOT a feature
- Retinal venous leakage presenting as a haemorrhagic retinopathy which resolves with steroids
- Retinal neovascularisation +/- closure





Inflammation of veins most frequent when disease isolated to eye

• Intermediate uveitis - florid vitritis, leaky retinal vessels

• Ischaemic retinal vasculitis – mild vitritis

34 year old woman with recalcitrant intermediate uveitis – no associated systemic disease











### Ischaemic retinal vasculitis



Retinal neovascularisation can occur secondary to capillary closure or merely as a result of inflammation and hence is not helpful diagnostically Multiple sclerosis – with marked closure

Behcets – in absence of closure





Retinal capillaries are very rarely affected on their own in the absence of post capillary venular involvement

Syphilis, Whipple's disease

### Retinal vascular change in uveitis - diagnosis

Vessel affected	Disease	Clinical signs	FFA signs
<u>Arterioles</u>	Systemic vasculitis	CWS	Closure
	(SLE, PAN, Wegener's)	Occlusion	NVs rare
		Uveitis rare	
	Viral/protozoal infection (ARN, CNV, <i>Toxoplasm</i> a)	Peri-arteriolar infiltration	Arteriolar leakage
		Closure	Occlusion
<u>Capillaries</u>	Syphilis	Haem, exudate,	Masking
		microaneurysms	Focal leakage

### Retinal vascular change in uveitis - diagnosis

Vessel	Disease	Clinical signs	FFA features
affected	Behcets	Infiltrates, vein occlusions, CME,	Masking, leakage,closure
<u>Veins</u> -	Sarcoid	Nvs Periphlebitis, peripheral closure,	Focal leakage, closure, CME,
	Multiple sclerosis	NVs CME Vein sheathing, CME Venular sheathing	NVs Variable staining
	HLA B27/IBD Lymphoma Idiopathic	CME	Microvascular leakage, CME

### Conclusions

- The majority of "retinal vasculitis" affects retinal veins
- Retinal arterial disease with inflammation is usually due to viruses
- Retinal arterial disease without inflammation is usually due to systemic vasculitis and rarely causes uveitis