

# **IMPORTANT SYSTEMIC ASSOCIATIONS OF UVEITIS**

## **1. Spondylarthropathies**

## **2. Juvenile idiopathic arthritis**

## **3. Sarcoidosis**

- Systemic features
- Ocular features

## **4. Behçet disease**

- Systemic features
- Ocular features




## **5. Vogt-Koyanagi-Harada syndrome**

## **6. Inflammatory bowel disease**

- Ulcerative colitis
- Crohn disease

## **7. Tubulointerstitial nephritis and uveitis**

# Spondylarthropathies

|  |                               | <b>Gender</b>    | <b>HLA-B27</b> | <b>Acute iritis</b> |
|--|-------------------------------|------------------|----------------|---------------------|
|    | <b>Ankylosing spondylitis</b> | <b>70% males</b> | <b>95%</b>     | <b>30%</b>          |
|    | <b>Reiter syndrome</b>        | <b>90% males</b> | <b>60%</b>     | <b>20%</b>          |
|  | <b>Psoriatic arthritis</b>    | <b>equal</b>     | <b>30%</b>     | <b>10%</b>          |

# Spondylarthropathies

**Sacroiliitis**

**Peripheral arthritis**

**Bowel inflammation**



**Ankylosing spondylitis**

**100%**

**20%**

**Common**

**Reiter syndrome**

**60%**

**100%**

**Uncommon**

**Psoriatic arthritis**

**30%**

**100%**

**Occasional**

# Clinical features of Reiter syndrome



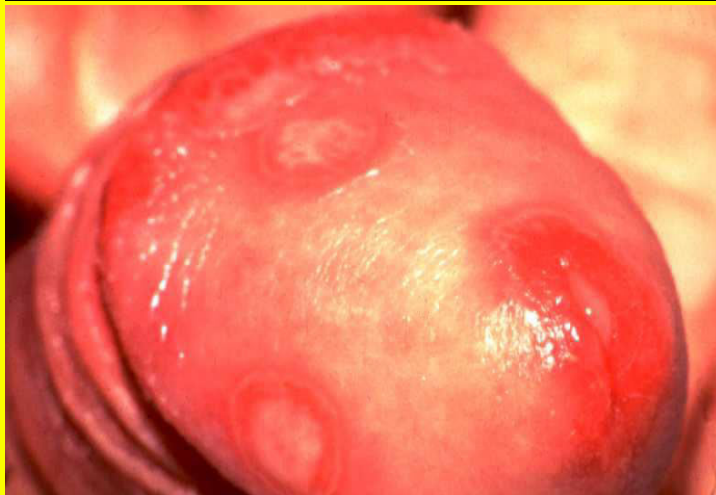
**Conjunctivitis**



**Plantar fasciitis**



**Painless oral ulceration**



**Urethritis and  
circinate balanitis**






**Keratoderma  
blenorrhagica**



**Nail dystrophy**

# Juvenile idiopathic arthritis

|                          | <b>Pauciarticular<br/>(60%)</b>   | <b>Polyarticular<br/>(20%)</b>  | <b>Systemic<br/>(20%)</b>   |
|--------------------------|---|---|---|
|                          |  |  |  |
| <b>Joints no.</b>        | <b>&lt; 5</b>   | <b>&gt; 4</b>   | <b>Variable</b>   |
| <b>Onset</b>             | <b>&lt; 6 years</b>   | <b>Variable</b>   | <b>Variable</b>   |
| <b>Systemic features</b> | <b>Absent</b>   | <b>Mild or absent</b>   | <b>Severe</b>   |
| <b>Positive ANA</b>      | <b>75%</b>  | <b>40%</b>  | <b>10%</b>  |
| <b>Iridocyclitis</b>     | <b>20%</b>  | <b>5%</b>   | <b>Absent</b>   |

# High risk factors for uveitis in JIA



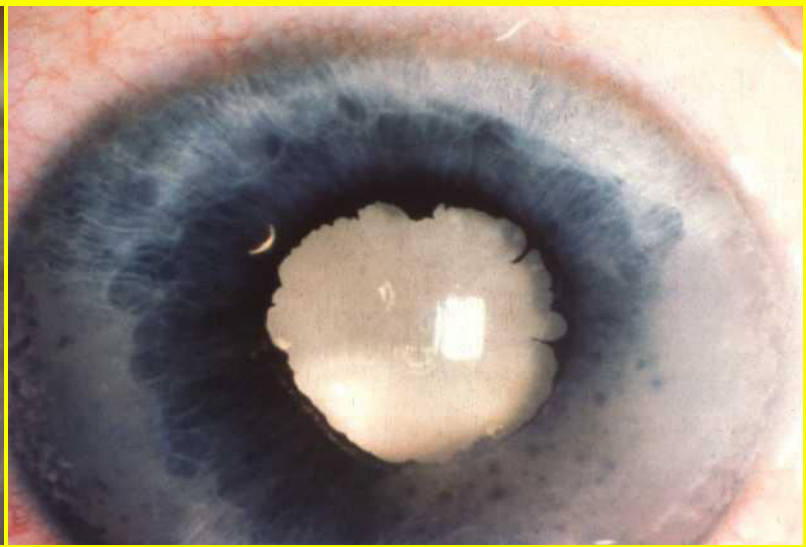
- **Girls**
- **Early onset**
- **Pauciarticular onset**
- **ANA**
- **HLA-DR5**



# Complications of uveitis



**Posterior synechiae - 30%**



**Cataract -20%**



**Glaucoma due to PAS - 15%**



**Band keratopathy - 10%**

# Systemic Features of Sarcoidosis

- 1. Idiopathic, multisystem non-caseating granuloma**
- 2. More common in blacks than whites**
- 3. Presentation**
  - Acute - third decade
  - Insidious - fifth decade
- 4. Organ involvement**
  - Lungs - 95%
  - Thoracic lymph nodes - 50%
  - Skin - 30%
  - Eyes - 30%



# Acute sarcoidosis



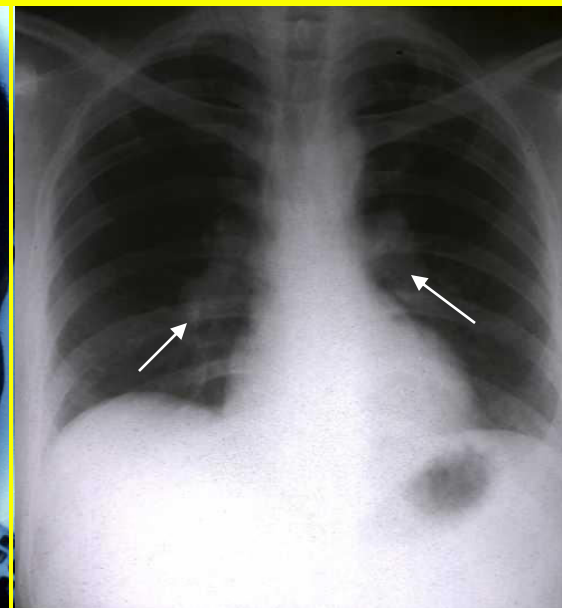
**Erythema nodosum**



**Parotid enlargement**

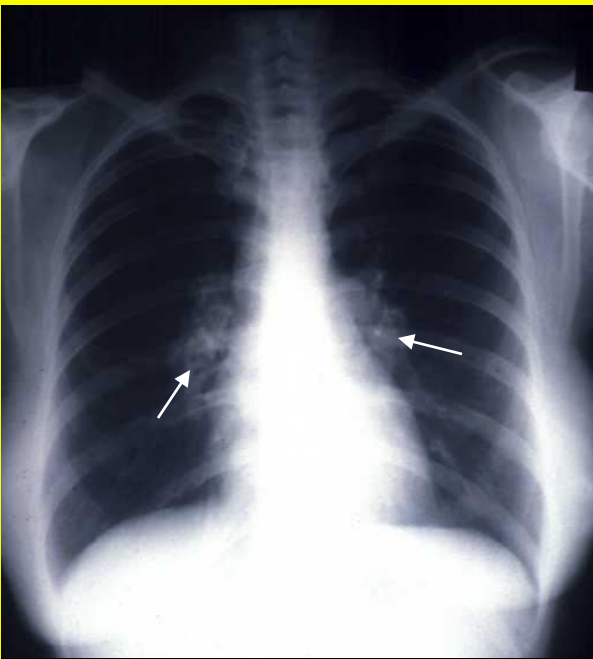


**Facial palsy**

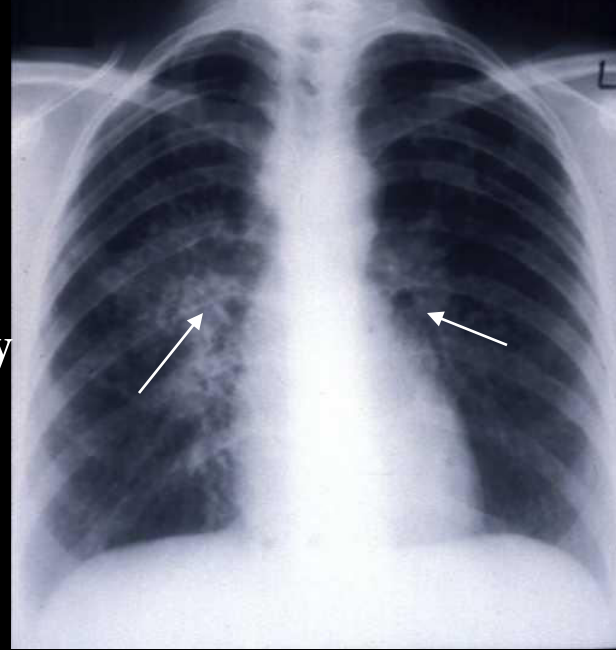


**Hilar lymphadenopathy**

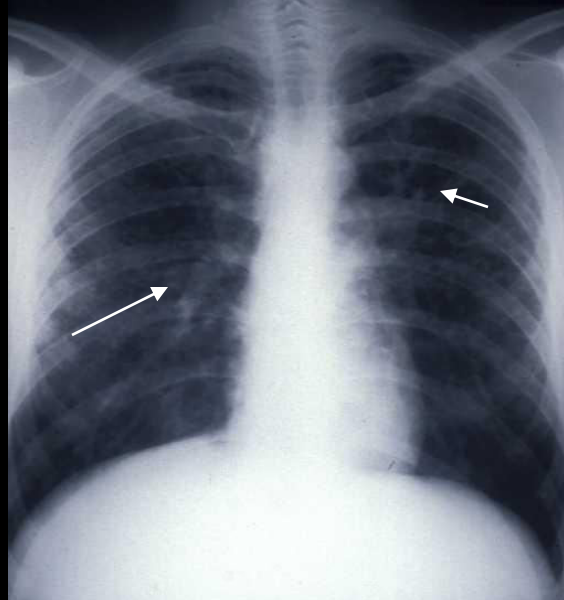
# Classification of sarcoid lung lesions



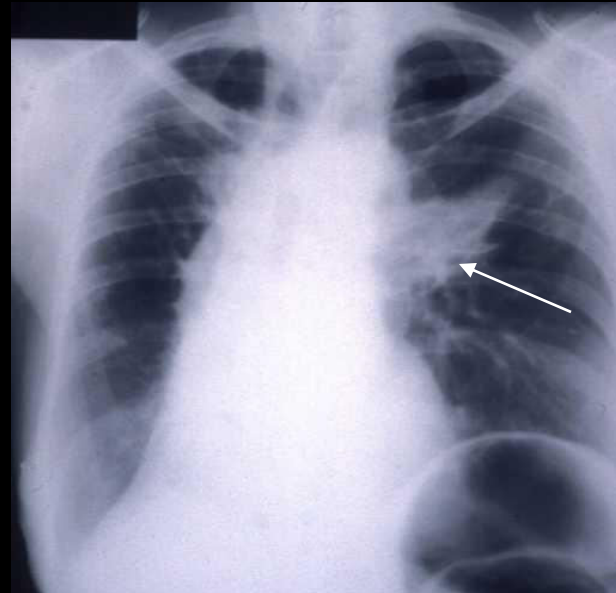
**Stage 1**  
Hilar  
Lymphadenopathy



**Stage 2**  
Hilar  
Lymphadenopathy  
and parenchymal  
infiltrates



**Stage 3**  
Parenchymal  
Infiltrates alone



**Stage 4**  
Fibrosis and  
Bronchiectasis

# Sarcoid skin lesions

**Granulomata**



**On face, buttocks and  
extremities**

**Lupus pernio**

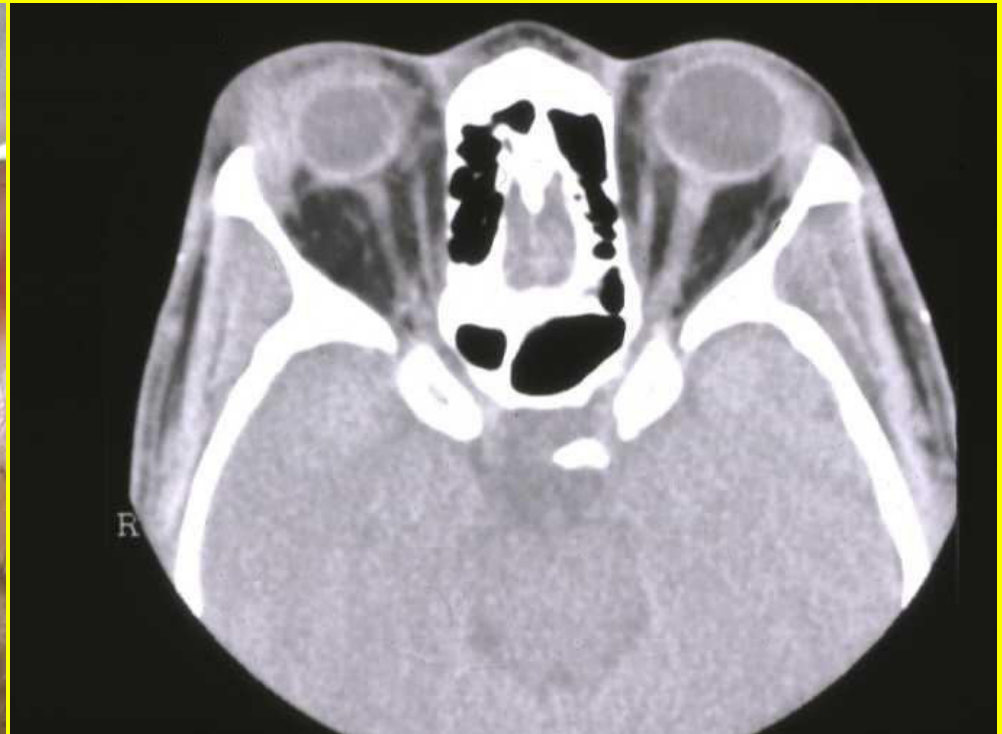


**Indurated, purple-blue lesions**

# Anterior segment lesions in sarcoidosis



**Conjunctival granuloma**



**Lacrimal gland involvement and dry eyes**



# Iridocyclitis in sarcoidosis

**Acute non-granulomatous**



**In young patients with acute sarcoid**

**Chronic granulomatous**



**In older patients with chronic sarcoid**



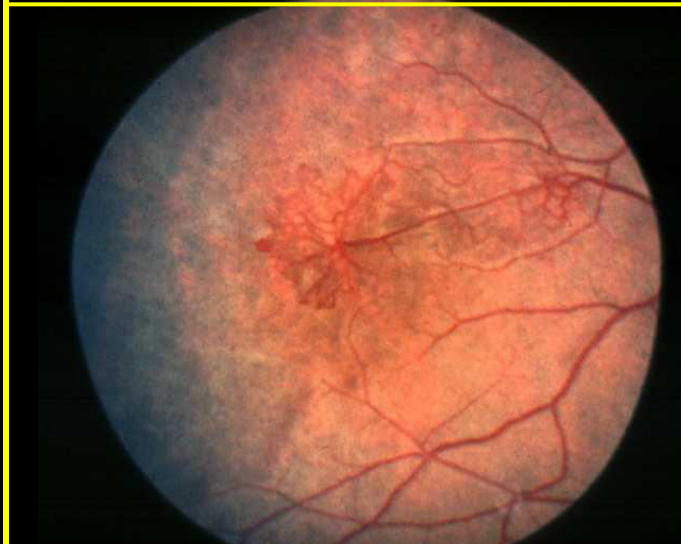
# Posterior segment lesions in sarcoidosis



**Subtle periphlebitis**



**Candlewax drippings**

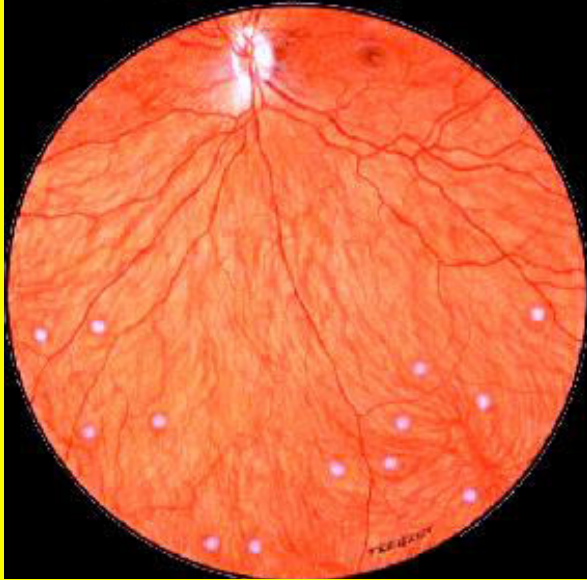


**Peripheral neovascularization**

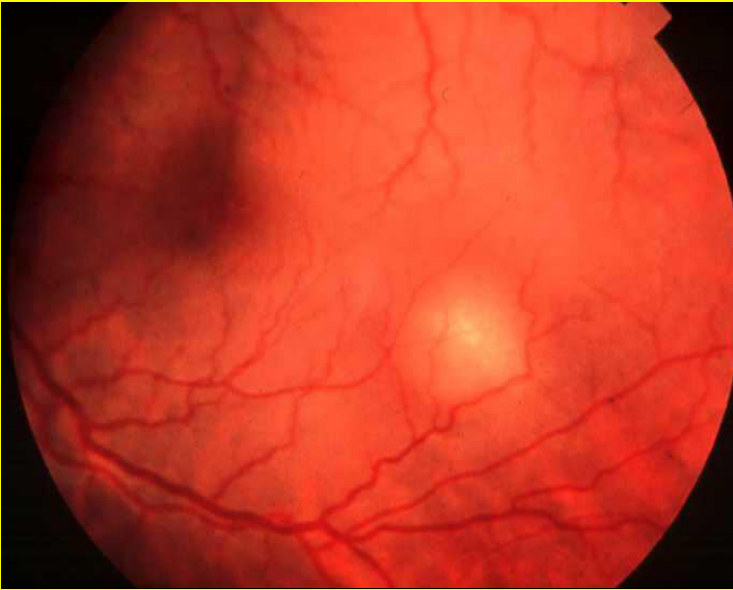


**Vitritis and snowballs**

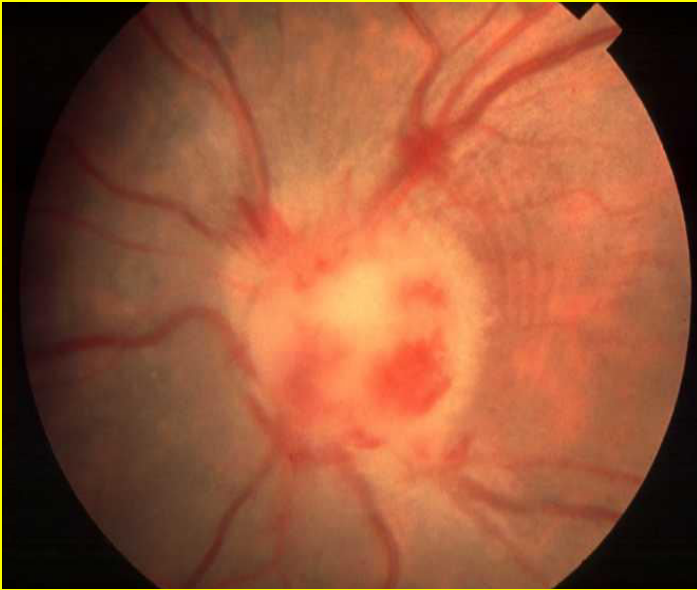
# Fundus granulomata in sarcoidosis



**Retinal and preretinal**



**Choroidal**



**Optic nerve head**

# Behçet Disease

- 1. Idiopathic multisystem disease**
- 2. Presentation - third to fourth decade**
- 3. Most prevalent in Mediterranean region and Japan**
- 4. Associated with HLA-B5 in Turkey and Japan**
- 5. Major diagnostic criteria**
  - Oral aphthous ulceration (100%)
  - Genital ulceration (90%)
  - Skin lesions (80%)
  - Uveitis (70%)

# Mucocutaneous ulceration in Behçet disease



**Oral aphthous ulceration - painful, recurrent**



**Genital ulceration**



# Skin lesions in Behçet disease



**Erythema nodosum**



**Acneiform**



**Pustule after scratching skin  
(pathergy test)**



**Lines after stroking skin  
(dermatographism)**



# Vascular lesions in Behçet disease

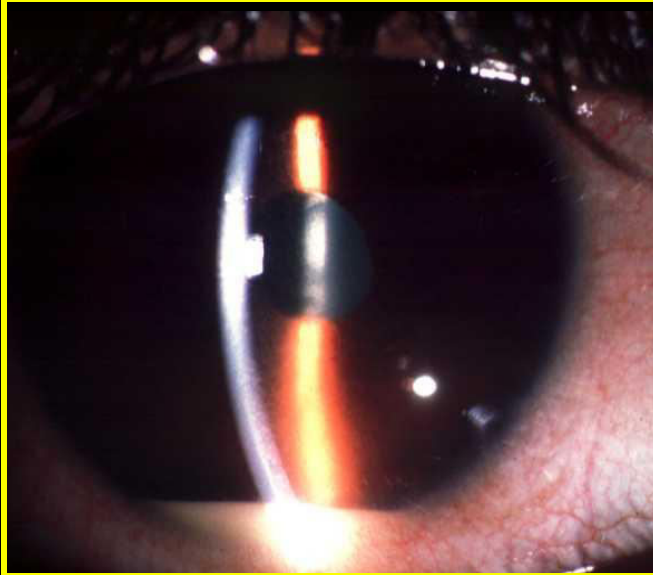


**Migratory thrombophlebitis  
of extremities**

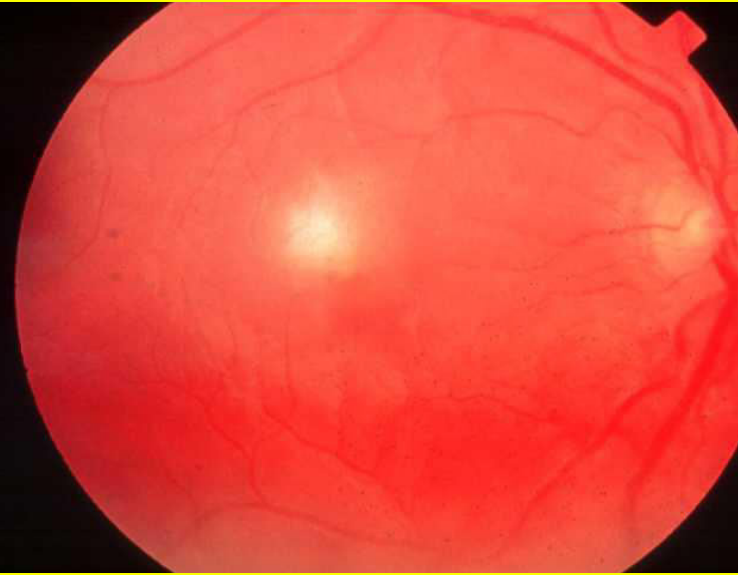


**Obliterative thrombophlebitis  
of major internal veins**

# Uveitis in Behçet disease



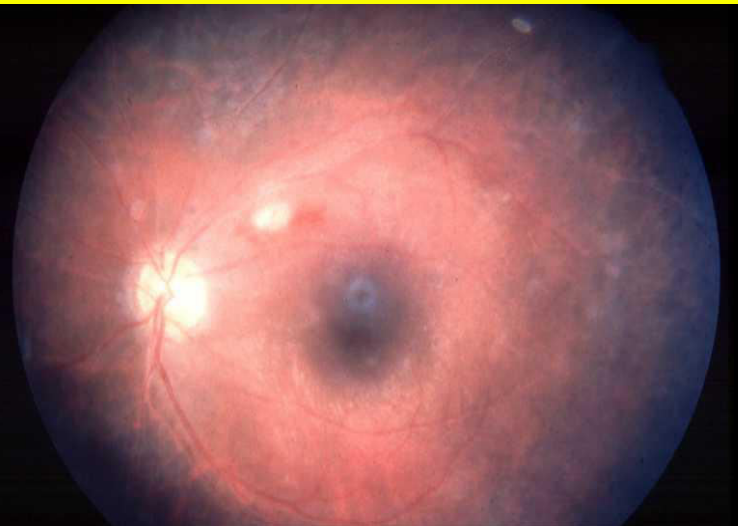
**Acute iritis**



**Retinitis**

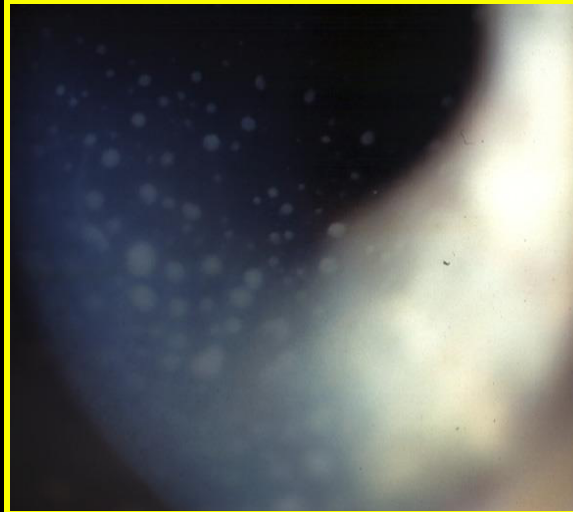


**Occlusive periphlebitis**



**Diffuse leakage**

# Signs of Vogt-Koyanagi syndrome



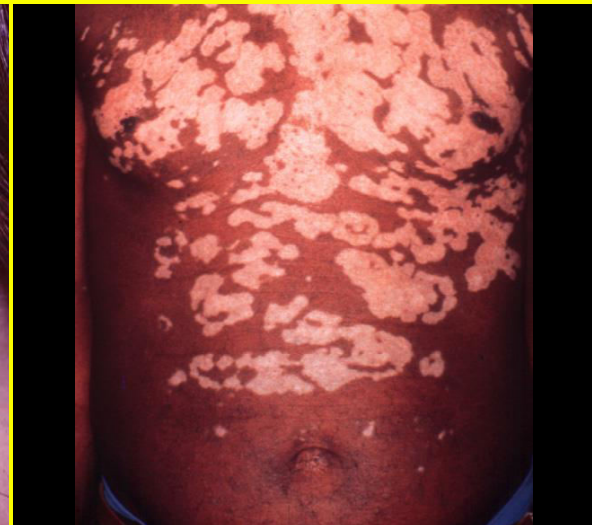
**Granulomatous  
iridocyclitis**



**Alopecia**



**Poliosis**



**Vitiligo**



# Harada syndrome

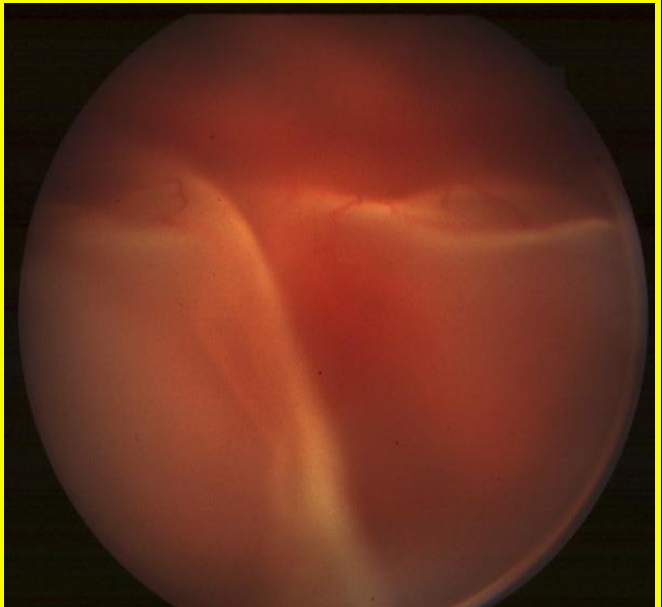
## Progression of Harada disease



**Multifocal choroiditis**



**Multifocal sensory retinal detachments**



**Exudative retinal detachment**

# Inflammatory bowel disease

## Ulcerative colitis



- Large bowel ulceration
- Acute iritis - uncommon

## Crohn disease

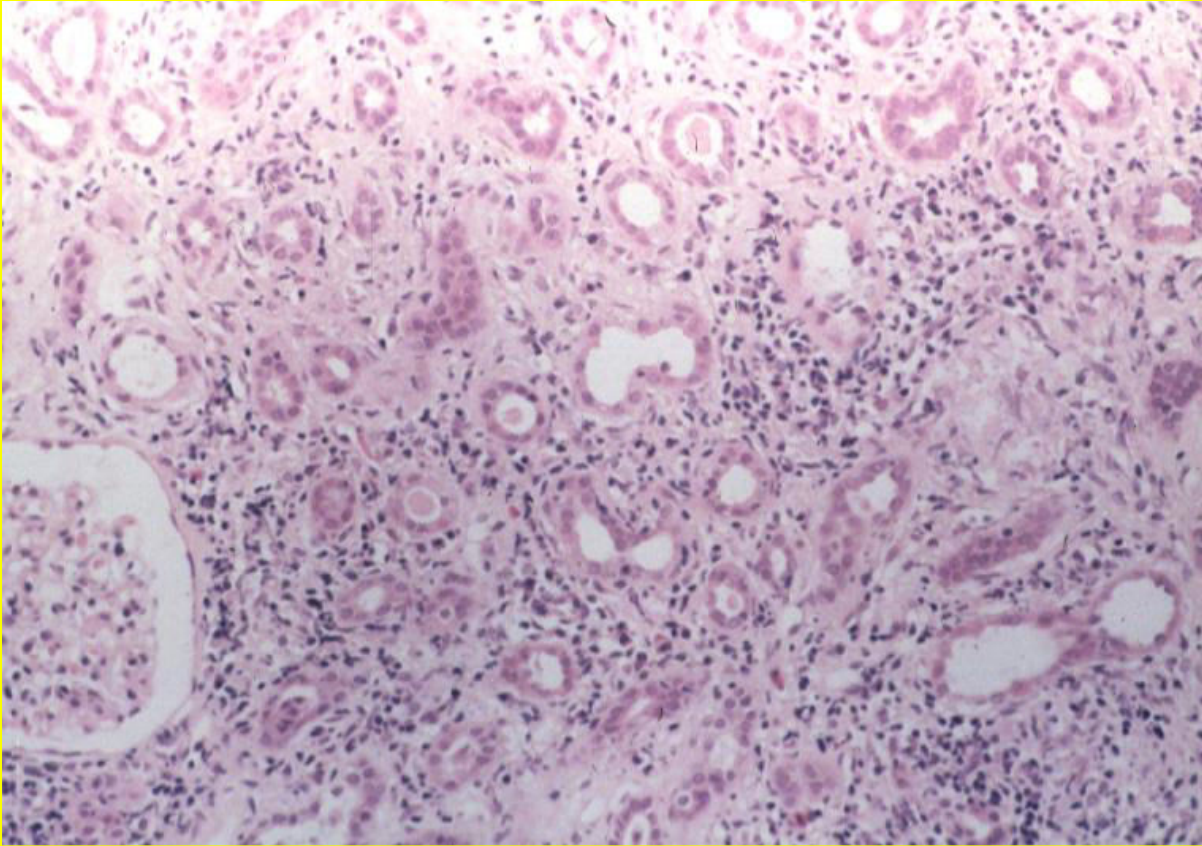


- Stricture and 'rose thorn' ulceration
- Acute iritis - uncommon



# Tubulointerstitial nephritis and uveitis (TINU)

## Renal histology



- Most frequently affects women and children
- Hypersensitivity reaction to drugs
- Bilateral, recurrent anterior uveitis

## Urine



- Proteinuria and renal failure
- Good response to systemic steroids