

GRANULOMATOUS AND VASCULITIC CONDITIONS

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Granulomatous and Vasculitic Conditions

- Heterogenous group of Chronic inflammatory disorders**
- Primary or Secondary**
- Have overlap of clinical manifestations**
- Can all have serious consequences**

Granulomatous and Vasculitic Conditions

Unknown aetiology

Sarcoidosis
Wegener's
Churge Strauss
Polyarteritis nodosa
SLE
Behçet's

Infective causes

Fungal disease
TB
Syphilis
Leprosy

Occupational

Berylliosis

Sarcoidosis and WG

- Clinical features – how to recognize?
- Investigation – what tests?
- Current concepts in management

Vasculitis screen

Blood tests:

ESR & CRP

Full blood count

Biochemistry, Calcium

Syphilis serology

ACE

ANCA

Biopsy

Chest X ray

Urinalysis:

blood / protein

24 hour calcium

SARCOIDOSIS

Clinical features

➤ Young adults women:men 2:1

➤ Multisystem disease

**Upper respiratory tract / salivary glands /
orbits / ears**

Chest

CNS / Joints / Heart / Skin / Kidneys

SARCOIDOSIS: pathogenesis

- Immune-mediated multi-organ disorder
- Non-caseating granulomata
- Unknown origin
- T-helper 1 – type response of T cells

SARCOIDOSIS: Sino-Nasal disease

Clinical Complaints

- Nasal obstruction / nose bleeds / crusting Anosmia
- Sinusitis
- Facial pain & swelling

Examination

- Large Inferior turbinate
- Septum: friable mucosa / nodules
Septal perforation
- Saddle deformity

SARCOIDOSIS: Skin conditions

- Erythema nodosum
- Lupus pernio



SARCOIDOSIS: ACE

Angiotensin Converting Enzyme

- Elevated in 50 -60% patients with sarcoidosis
- Elevated in articular disorders with other systemic disease
- Reduced by ACE inhibitors

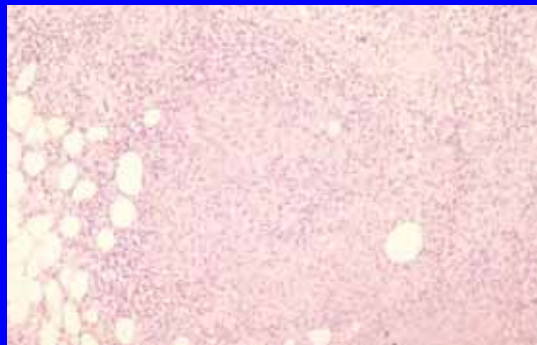
SARCOIDOSIS: ACE

- Originates in the lung
- Reflects the 'granuloma load'
- Used to monitor disease state

SARCOIDOSIS: Biopsy

Well-defined non-caseating granulomas

Nasal biopsy
Neck nodes
Oral mucosa
Conjunctiva



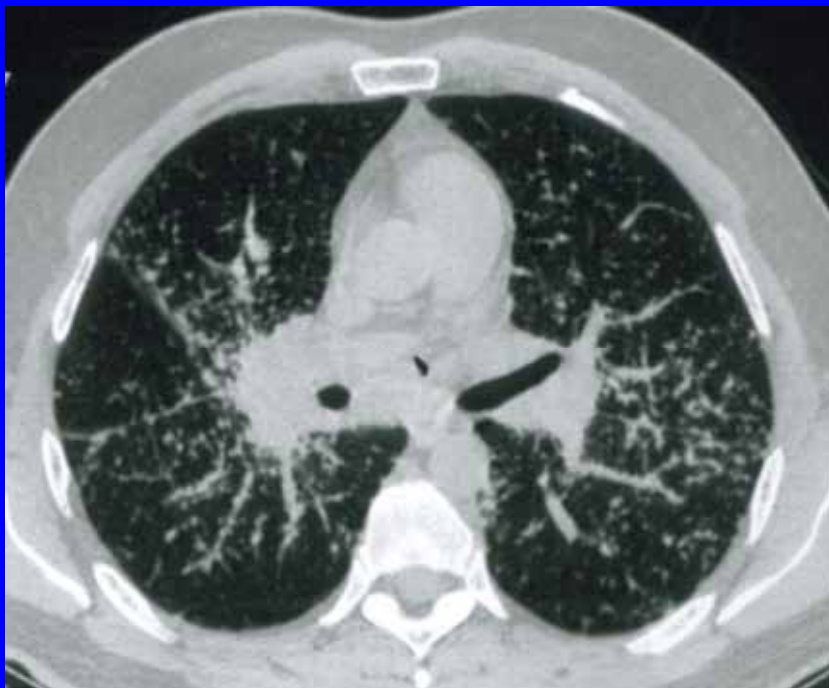
Transbronchial biopsy

can be positive even from macroscopically normal mucosa.

Sarcoidosis: Chest X ray



SARCOIDOSIS: High Resolution CT



Sino-nasal Sarcoidosis: Management

Topical Treatment: **Steroids sprays**
Saline douches
Glucose-glycerine drops

Surgery: **FESS for sinusitis**

Avoid / defer **Septal surgery**
Reconstructive surgery

Spontaneous resolution can occur

SARCOIDOSIS Medical Management

Systemic steroids - ? duration

Steroid limiting strategy

- Methotrexate
- Anti-TNF alpha blocker (Infliximab)

Wegener's Granulomatous: WG

Patients fall into 2 categories

- 1 upper airway
- 2 affecting systems below upper airway

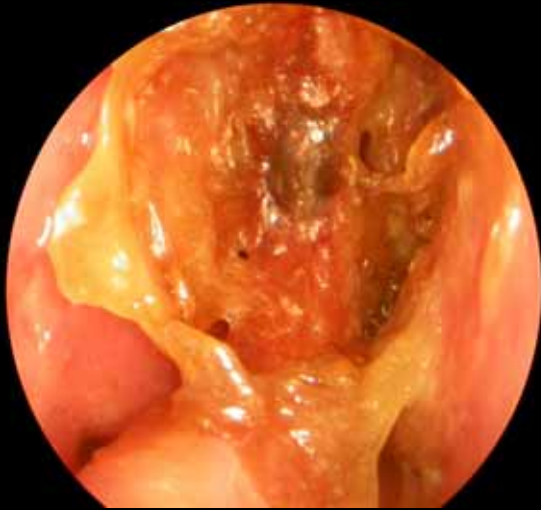
Upper airway WG

is likely to progress eventually to involve major organs below the neck.

WG Clinical Features



WG **Clinical features**



Nose



Ear

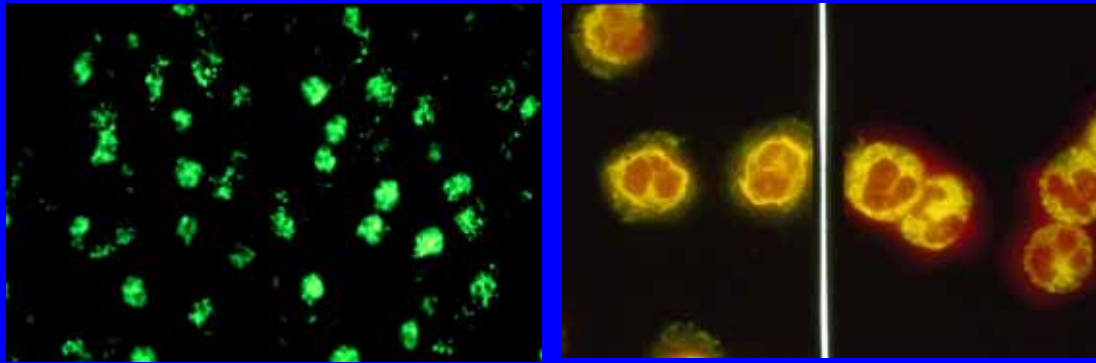
WG: Clinical features

Lacrimal obstruction
Scleritis / Uveitis
Pseudotumour

**Laryngeal and
pharyngeal disease**



WG: Anti-neutrophil cytoplasmic antibodies



- indirect immunofluorescence
- quantify target antigens with ELISA

WG: Anti-neutrophil cytoplasmic antibodies

pANCA: *perinuclear*

MPO: Myeloperoxidase

cANCA: *cytoplasmic*

PR3: Serine Proteinase 3

WG: cANCA

- Very specific and relatively sensitive
 - Poor screening tool
Good for diagnostic confirmation
 - PR3 present in 90% of patients
- may be negative!
-

- Does not always follow disease activity

Wegener's Granulomatosis Biopsy sites

Upper airway

- Often shows 'non-specific inflammation'
- Biopsy only abnormal looking mucosa
- Substantial and deep

Renal biopsy: more reliable.

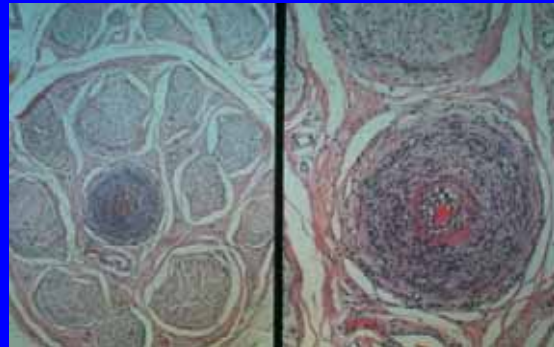
Lung biopsy: if diagnosis uncertain.

WG: Biopsy

Stains: H&E
Immunofluorescence

Ideal Positive result:

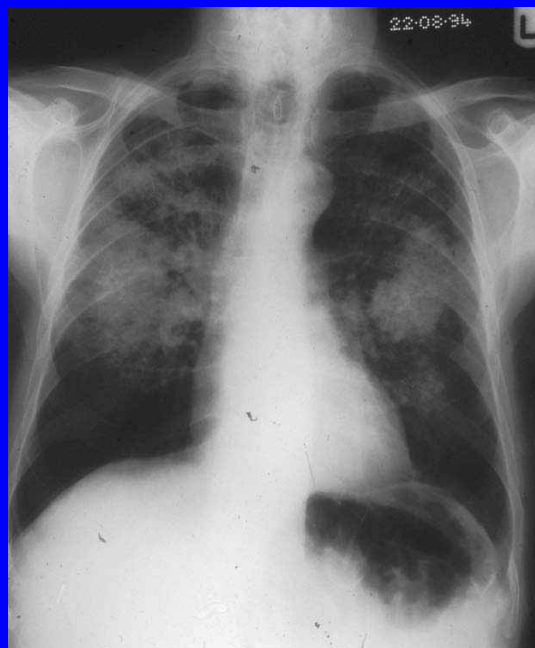
- 1 Vasculitis
- 2 Granulomas
- 3 Necrosis



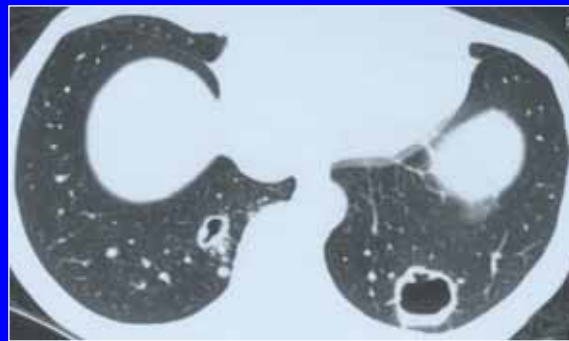
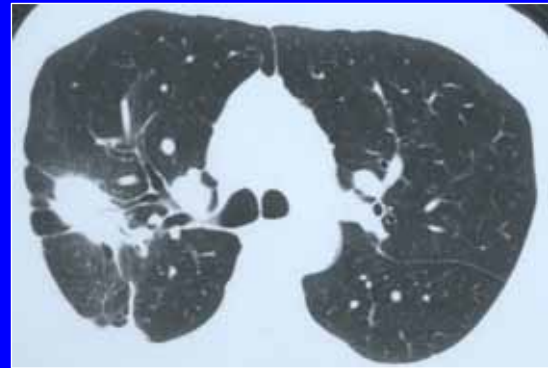
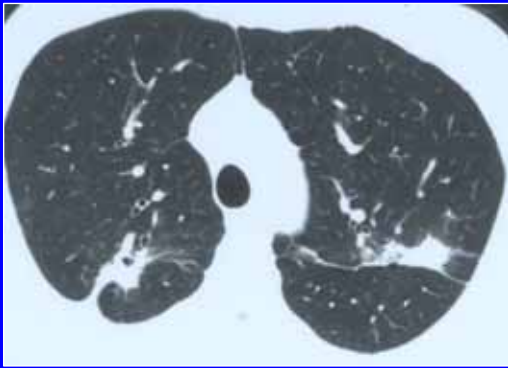
Histological confirmation

Important to obtain before treatment

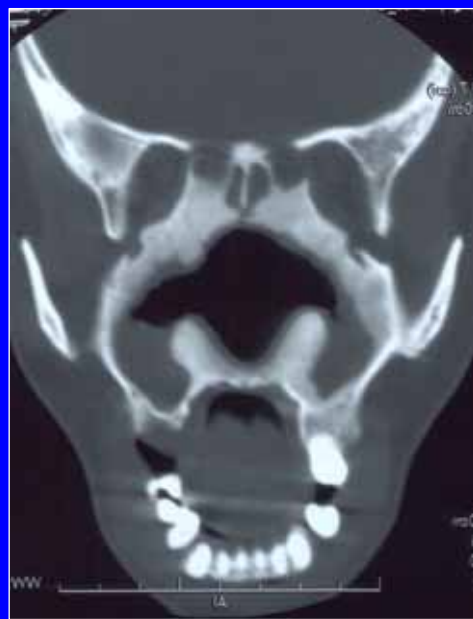
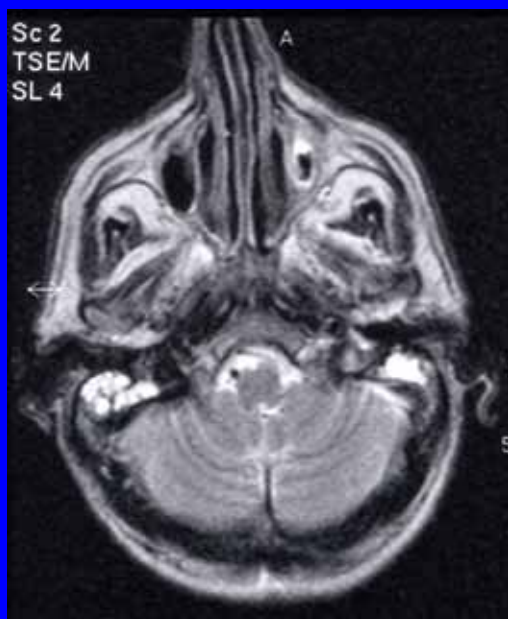
WG: Chest X ray



WG: High Resolution CT



WG: Imaging



WG - imaging



WG: Clinical features

Eyes



Nasal cavities



WG: Clinical features



WG and surgery



WG and Surgery Nasal Reconstruction

- ? Wait for several years in remission.
- Most important factor is to be in remission
- How to recognize a Relapse or Flare-up ?
 - Risk decreases with time
 - Signs: Clinical features most important
 - Surgery does **not** induce flare-up

WG: Medical Management

- Aim to induce and maintain remission
 - low-grade grumbling disease
 - immunosuppression
- Risk of relapse
- Dilemma - when to stop treatment

WG: Cytotoxic therapy & Systemic steroids

A Upper airway

Methotrexate

High dose Septrin Forte

B Uncertain diagnosis

Methotrexate / monitor response

C High clinical activity

Cyclophosphamide - toxic

Monoclonal antibody (Rituximab)

WG and Cocaine



WG and Cocaine

