Rheumatology in ICU

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Rheumatic diseases

More Common

- SLE
- RA
- Scleroderma
- Dermatomyositis / polymyositis
- Gout

Uncommon

- Vasculitis
- Wegener’s granulomatosis
- Takayasu’s arteritis
- Relapsing polychondritis

- Most rheumatic disorders managed at outpatient
- 10% to 25% hospitalization
- < 10% require intensive care
- 20% first diagnosed rheumatic disease at ICU
- In singapore study: RA > SLE > scleroderma (total 75%)
- Organ involvement: Resp > renal > GI > nervous system
- Infection > 50%
- Mortality 42 – 50%
- Poor prognostic factors: renal failure, coma, acute abdomen

Annals of the Rhem Dis 1992;52:627-631
Critical Care Clinics 2002;18(4):729-48

Reasons for ICU admission

Related to underlying rheumatic disease

- Flare up
- Develop new, life-threatening manifestations
- Infections
- Adverse effects of drugs esp immunosuppressant
- Malignancy due to prolong use of cytotoxic drugs or association with underlying malignancy

NOT related to underlying rheumatic disease

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NOT related to underlying rheumatic disease

Diseases pattern in ICU

A) Known rheumatic disease
   - Disease exacerbation
   - Treatment complications
   - Infection

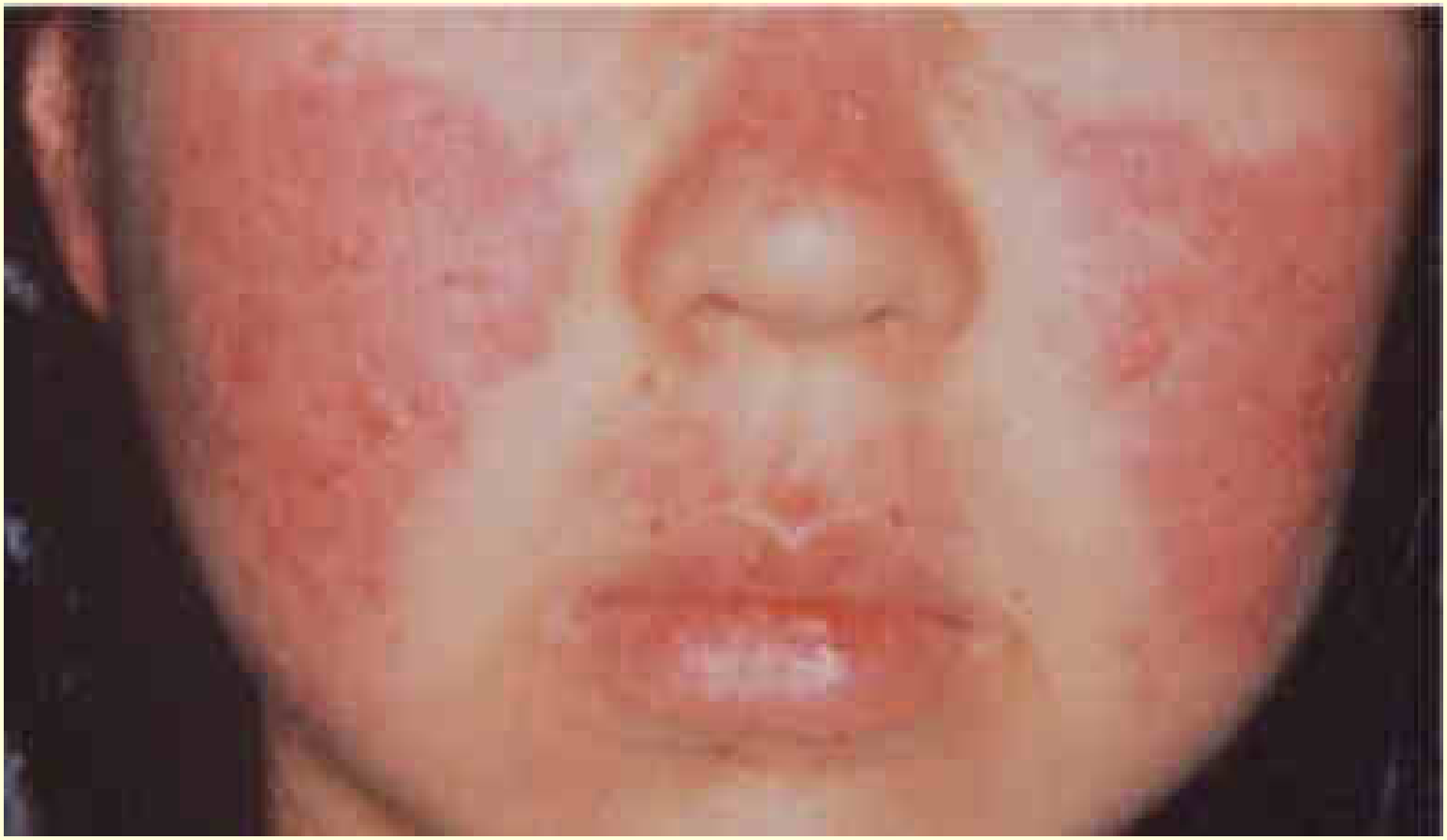
B) Unknown rheumatic disease presented with life threatening manifestation

c) Arthritis in critical patients

Common rheumatic diseases encountered in ICU

1. SLE
2. Rheumatoid Arthritis
3. Dermatomyositis / polymyositis
4. Scleroderma

Systemic lupus erythematosus (SLE)
系統性紅斑狼瘡



What is SLE?

- Systemic disease
- Characterized by autoantibodies directed against self-antigens, immune complex formation, and immune dysregulation
- Can damage to any organ especially kidney, skin, blood cells, and the CNS, .
- Clinical symptoms markedly variable
- may present with many years of symptoms or with acute life-threatening disease

SLE-epidemiology

- F:M 9:1
- Peak 15-40
- 40% , onset > 60 yrs old
- Prevalence rates are higher in Asian, Latin American, and black patients.

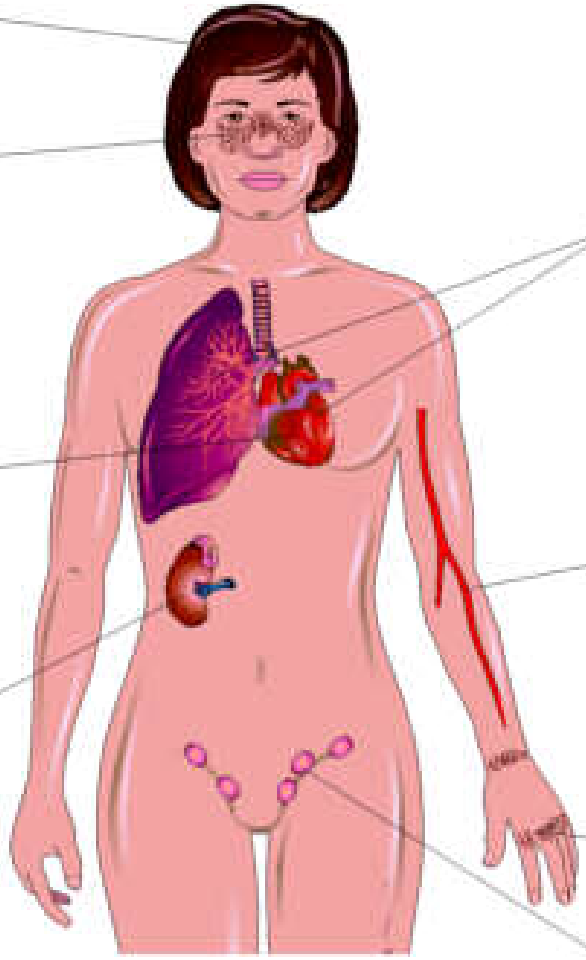
SLE

Seizures
Cerebral vasculitis

Butterfly rash
Discoid lupus
Oral ulceration
Hair loss

Pleuritis
Plerual effusion
Pulmonary haemorrhage
Lupus pneumonitis
Pulmonary hypertension

Glomerulonephritis
Renal failure



Endocarditis
Myocarditis
Pericarditis
Pericardial tamponade

Hemolytic anaemia
Leukopenia
Thrombocytopenia
TTP

Arthritis
Raynand's phenomenon

Lymphadenophathy

SLE – diagnostic criteria

1. Malar rash
2. Discoid rash
3. Photosensitivity
4. Oral ulcers
5. Arthritis
6. Serositis
7. Renal disorder
8. Neurological disorder
9. Haematologic disorder
10. Immunological disorder – DsDNA, anti-SM, anti-phospholipid antibodies
11. ANA +ve

the American College of Rheumatology → 4 out of 11 criteria are diagnostic of SLE

SLE -- pathogenesis

1) Genetic
2) Environmental
    - sunlight,
    - Stress
    - infectious agent
3) hormonal factors
4) Drug induced

It is likely that a combination of factors work together to cause the disease.

Treatment

- Minor symptoms
  - Rash – topical steroid, hydroxycholoquine
  - Arthritis – hydroxycholoquine, NSAID

- Major organ involvment
  - Prednisolone
  - Methotrexate
  - Azathioprine
  - Cyclophosphamide
  - Cyclosporine A
  - MMF (myclophenonate mofetil)
  - Tacrolimus
  - IVIg
  - Rituximab

Common presentations of SLE patients admitted to ICU

- confusion / convulsion
- renal failure
- infection

Approach to SLE with confusion / convulsion

DDx

- CNS infection
- Metabolic causes
  - Electrolyte disturbance
  - Uraemia
  - Posterior reversible encephalopathy syndrome
- Cerebral vascular accident
  - ischaemia
  - hemorrhage
- Cerebral vasculitis
- Cerebral sinuses thrombosis

History:

- Assess SLE disease activities
- Drug compliance

Physical examination:

- Oral ulcer, malar rash, arthritis, alopecia
- Weight loss

Investigation :

- Recent investigations
- CT brain
- LP
- EEG
- +/- MRI brain

Treatment:

- Underlying causes

Case 1

- F/20
- SLE with lupus nephritis since 14, given various immunosuppressants (Azathioprine, cyclosporine A and MMF)
- Disseminated TB 1/2005 (left knee, kidney), therefore immunosuppressant stopped
- Right eye CMV retinitis 2/2005

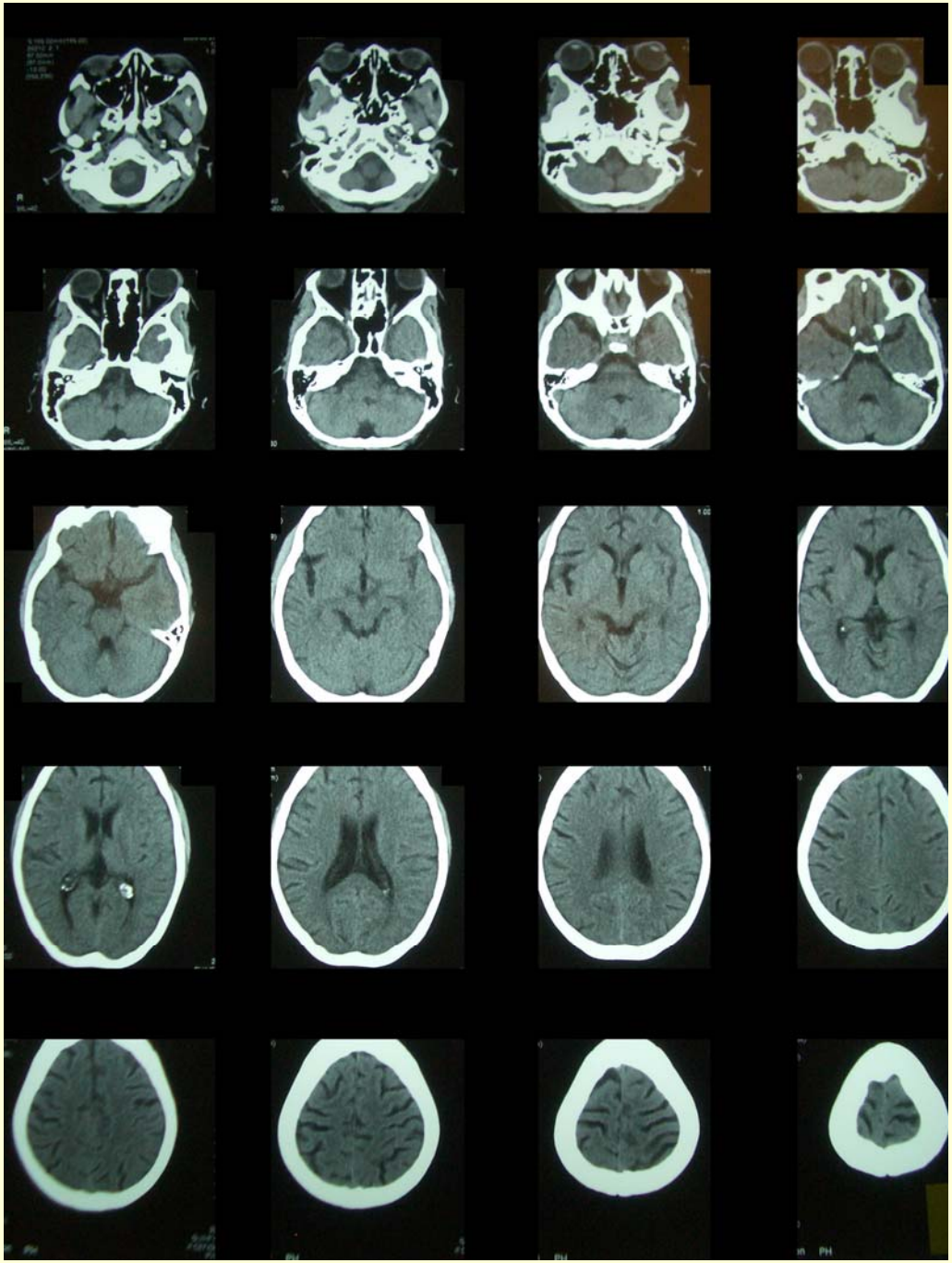
c/o

- headache 2/2005
- No vomiting, deny neck pain
- GCS 15/15

Ix:

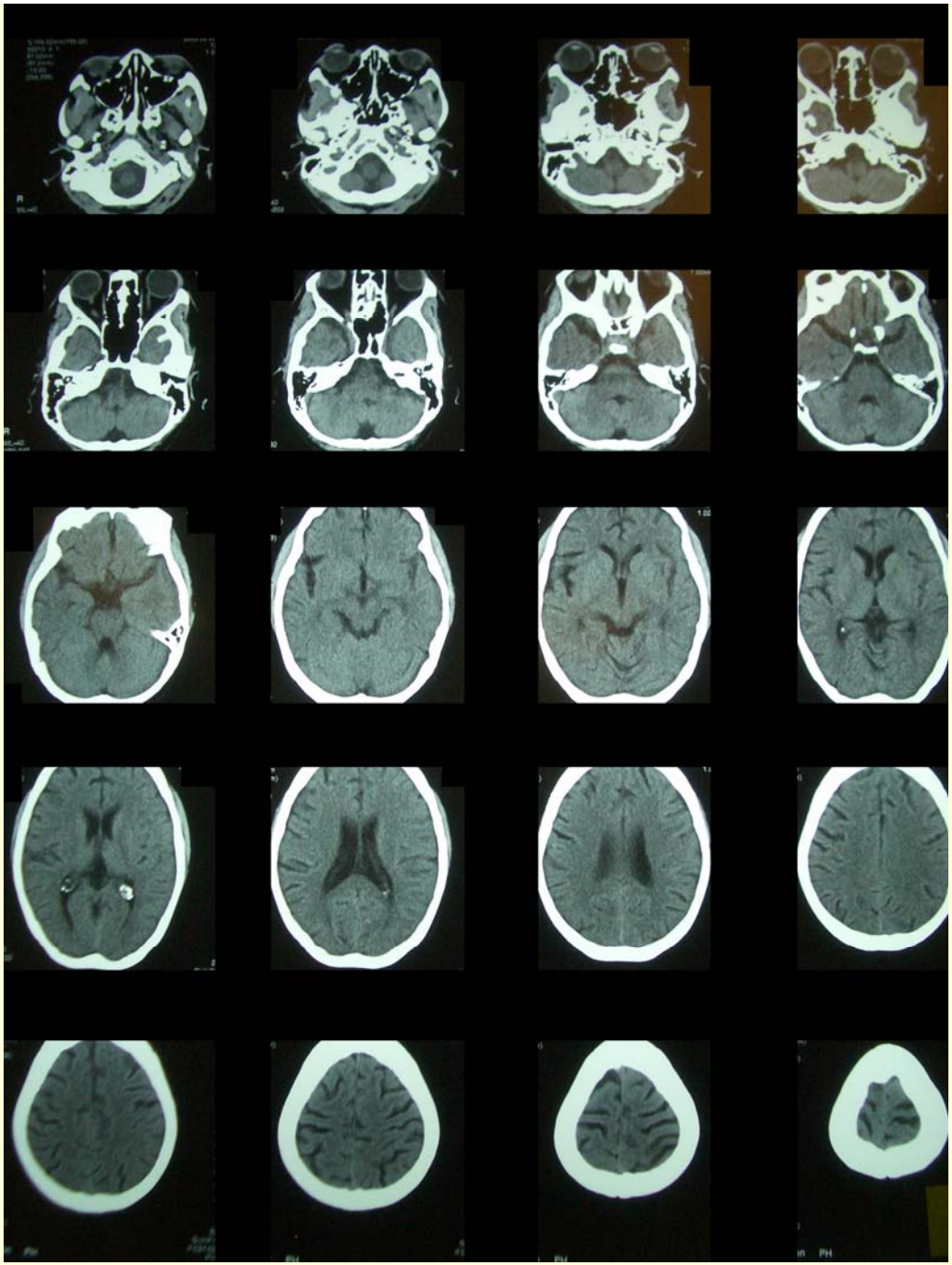
- Hb 9.8, plt 314, wcc/N/L 15.2/13.10.9
- Cr 95, alb 30
- DsDNA 31, C3 0.69 (static)
- ACA/LA –ve

CT brain

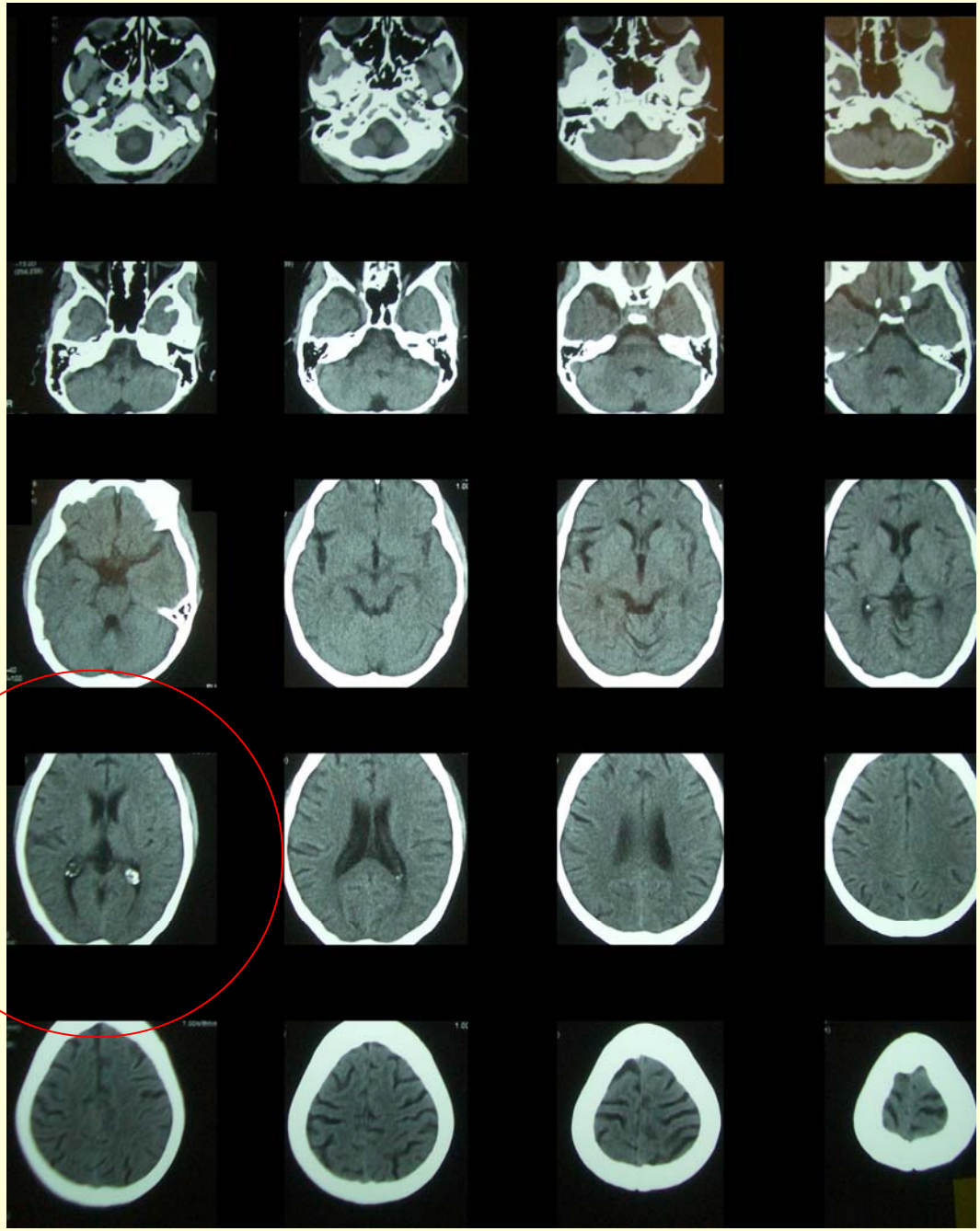


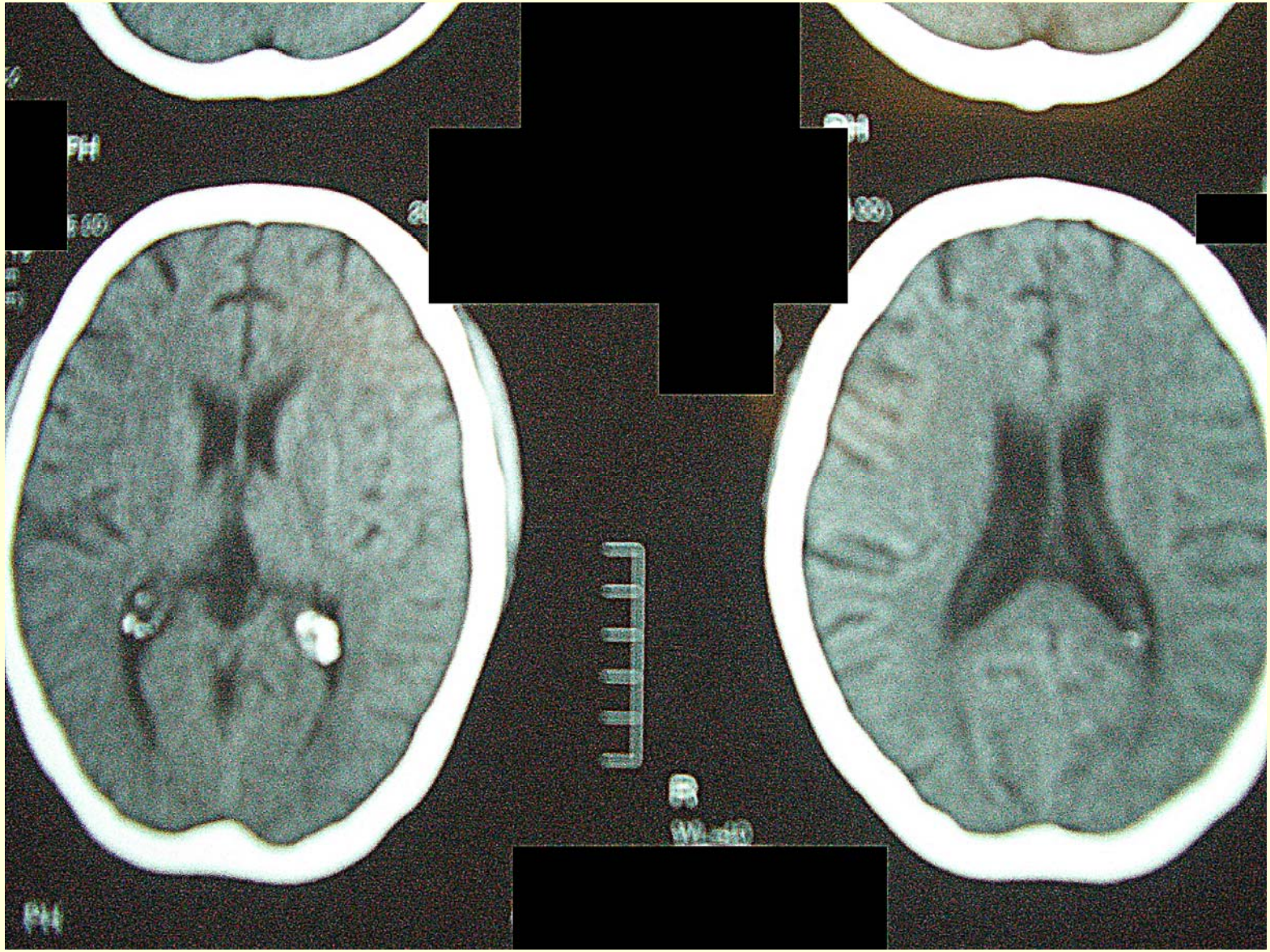
- On call MO : NAD
- Neurologist consulted as persistent headache

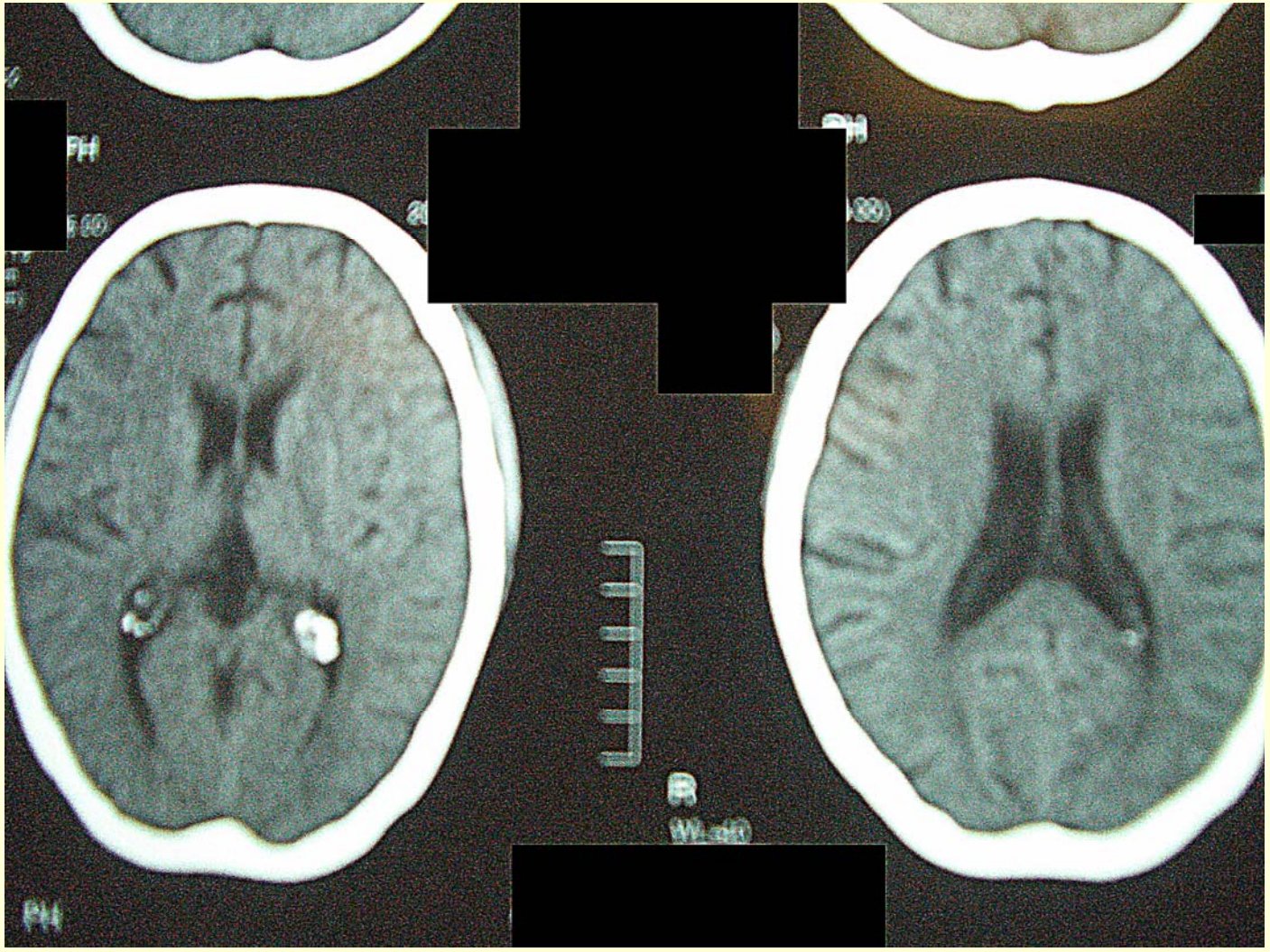
CT brain



CT brain





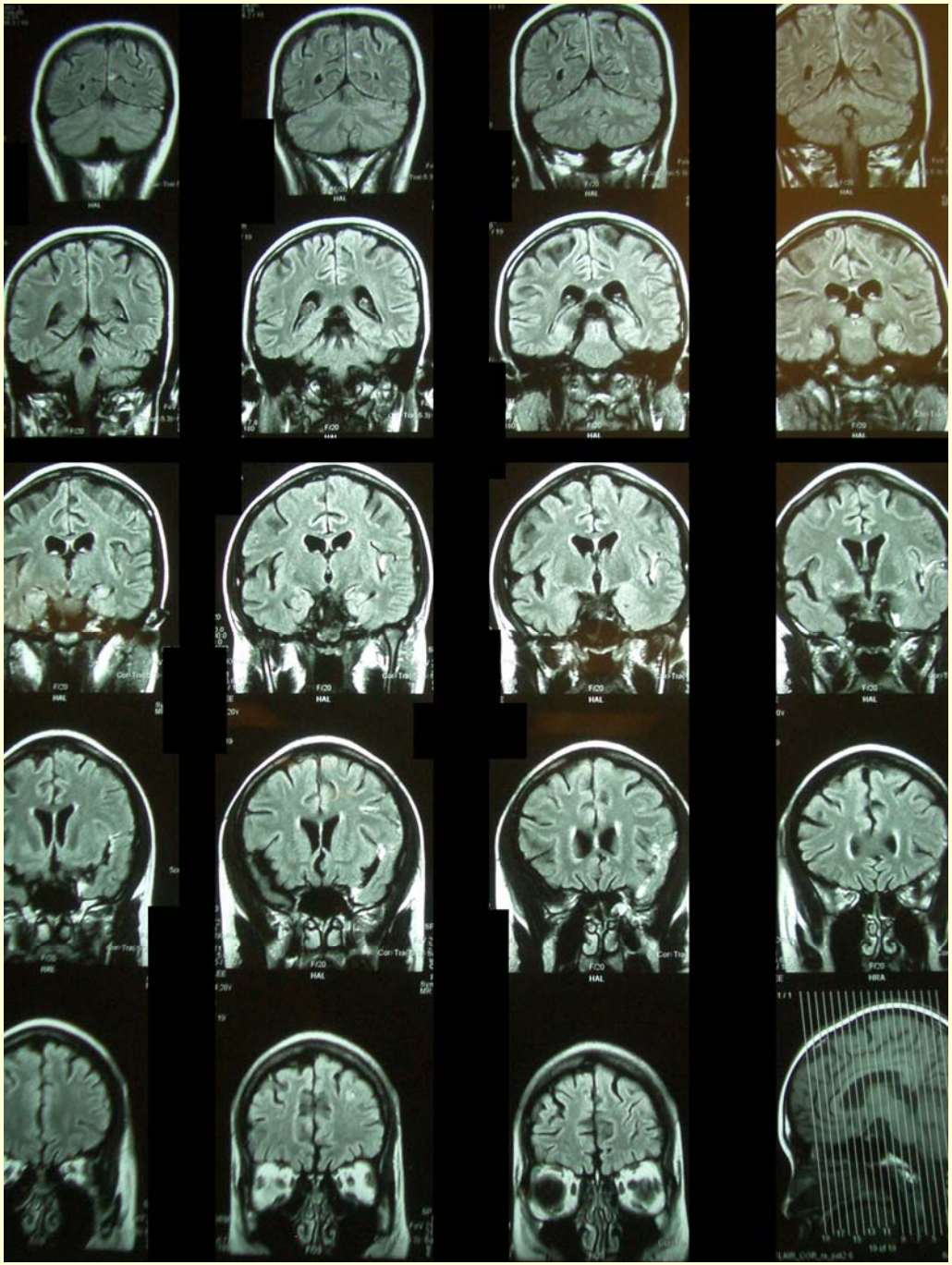


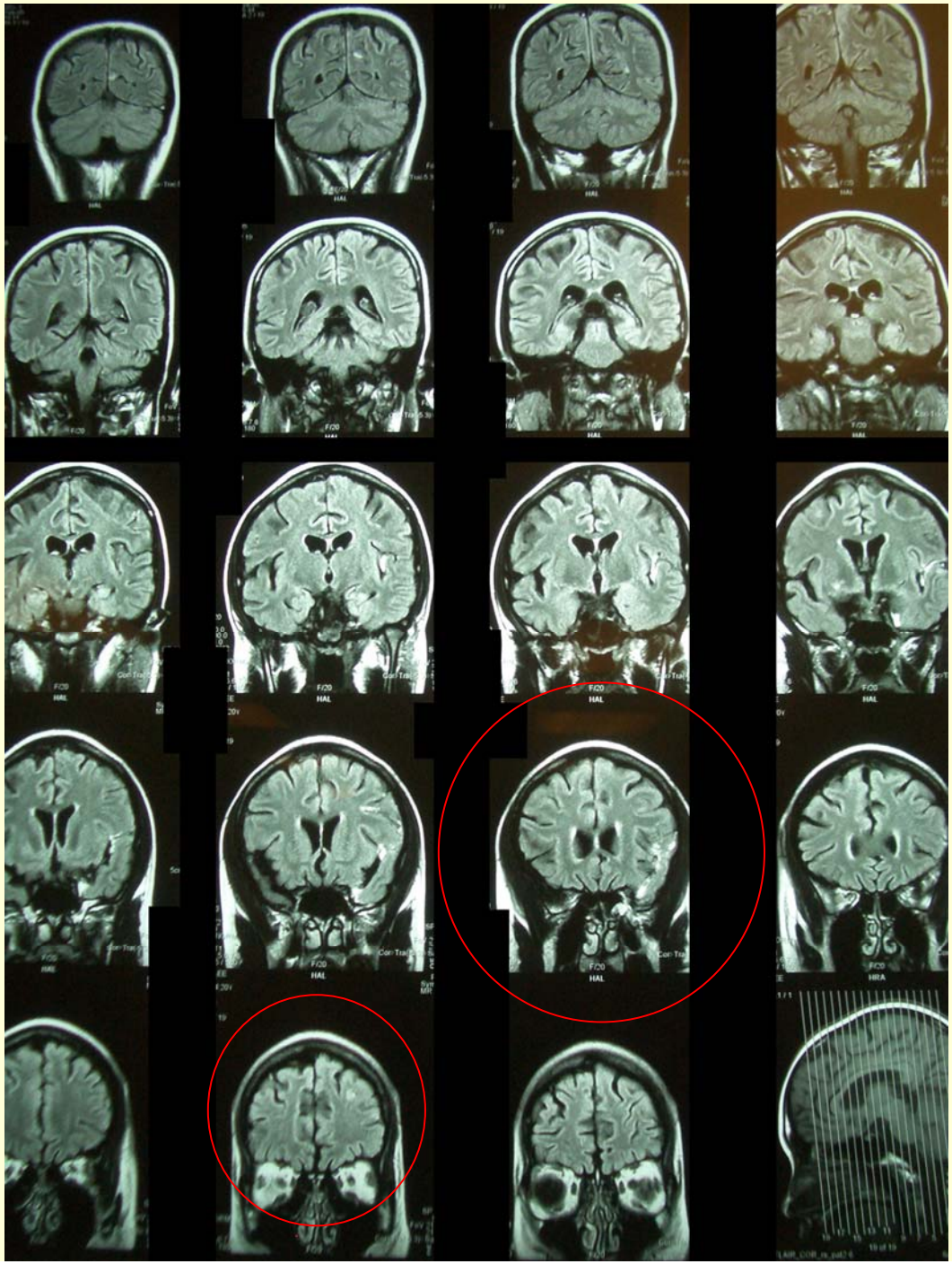
hydrocephalus over lateral, 3rd and 4 th ventricle

WHAT is the cause of hydrocephalus + headache ????

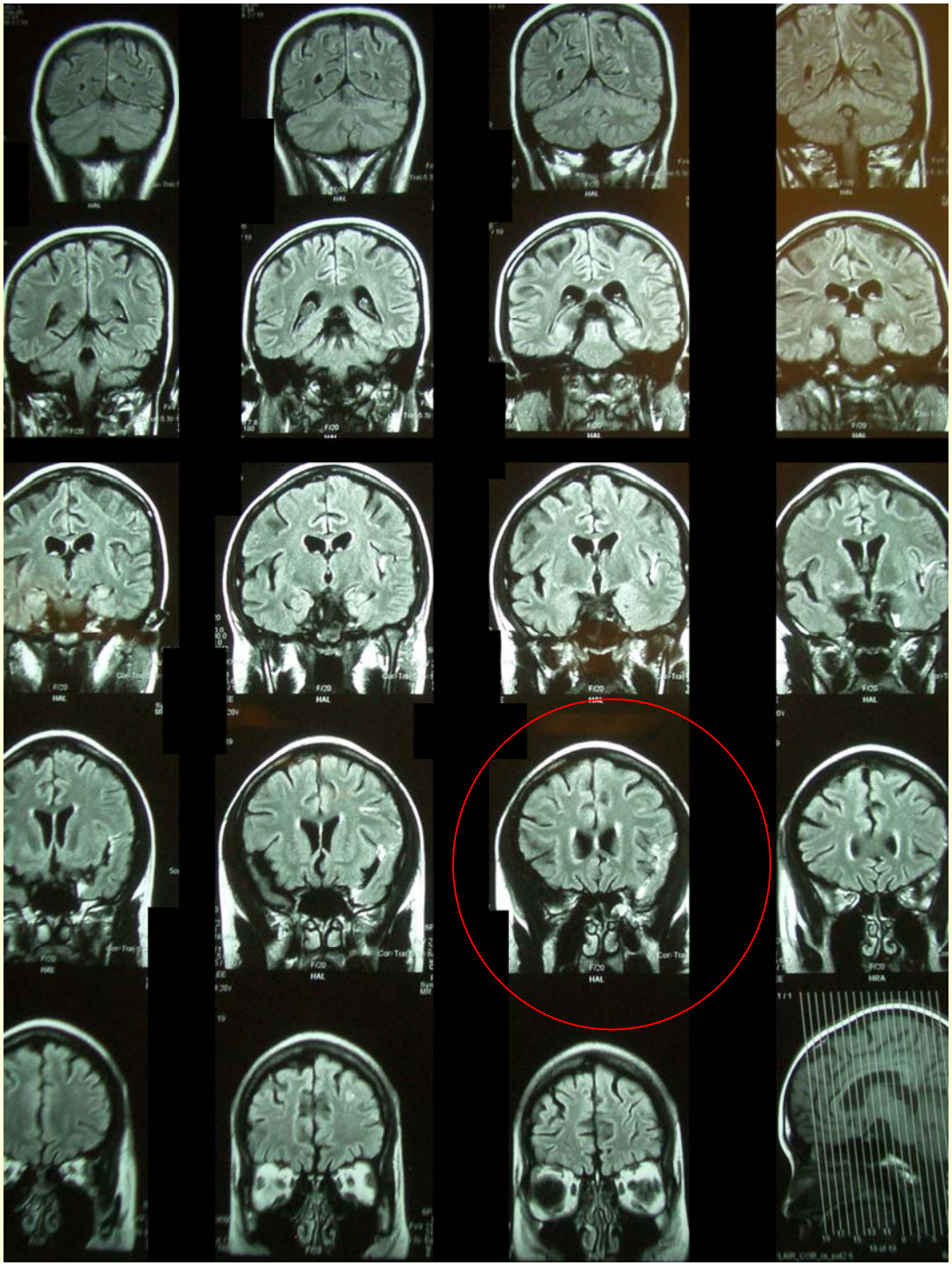
- ? sinus thrombosis
- ? TB meningitis
- ? SAH

- LP → exclude infection





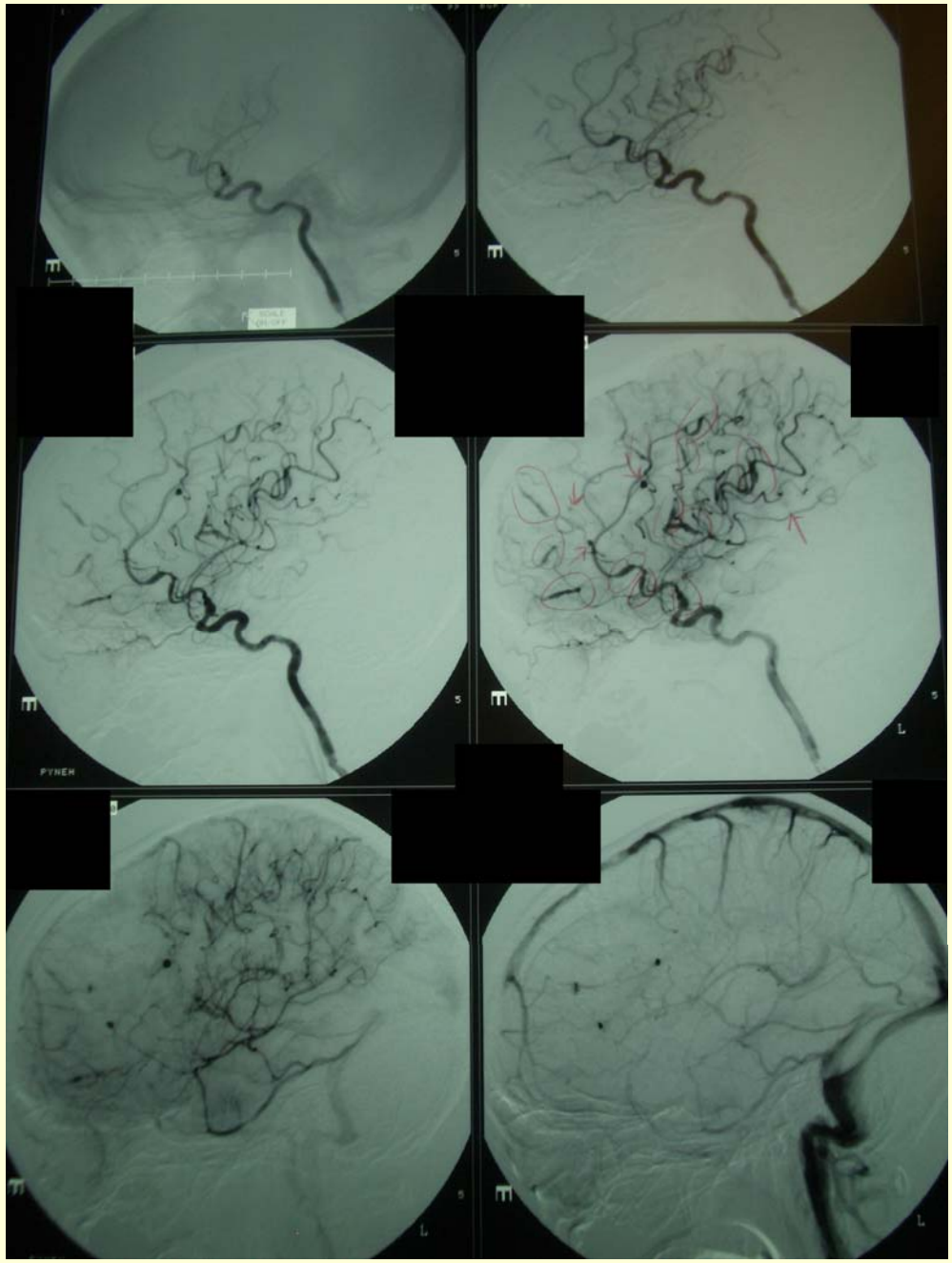
Subacute SAH in left sylvian fissure and medial aspect of left middle cranial fossa



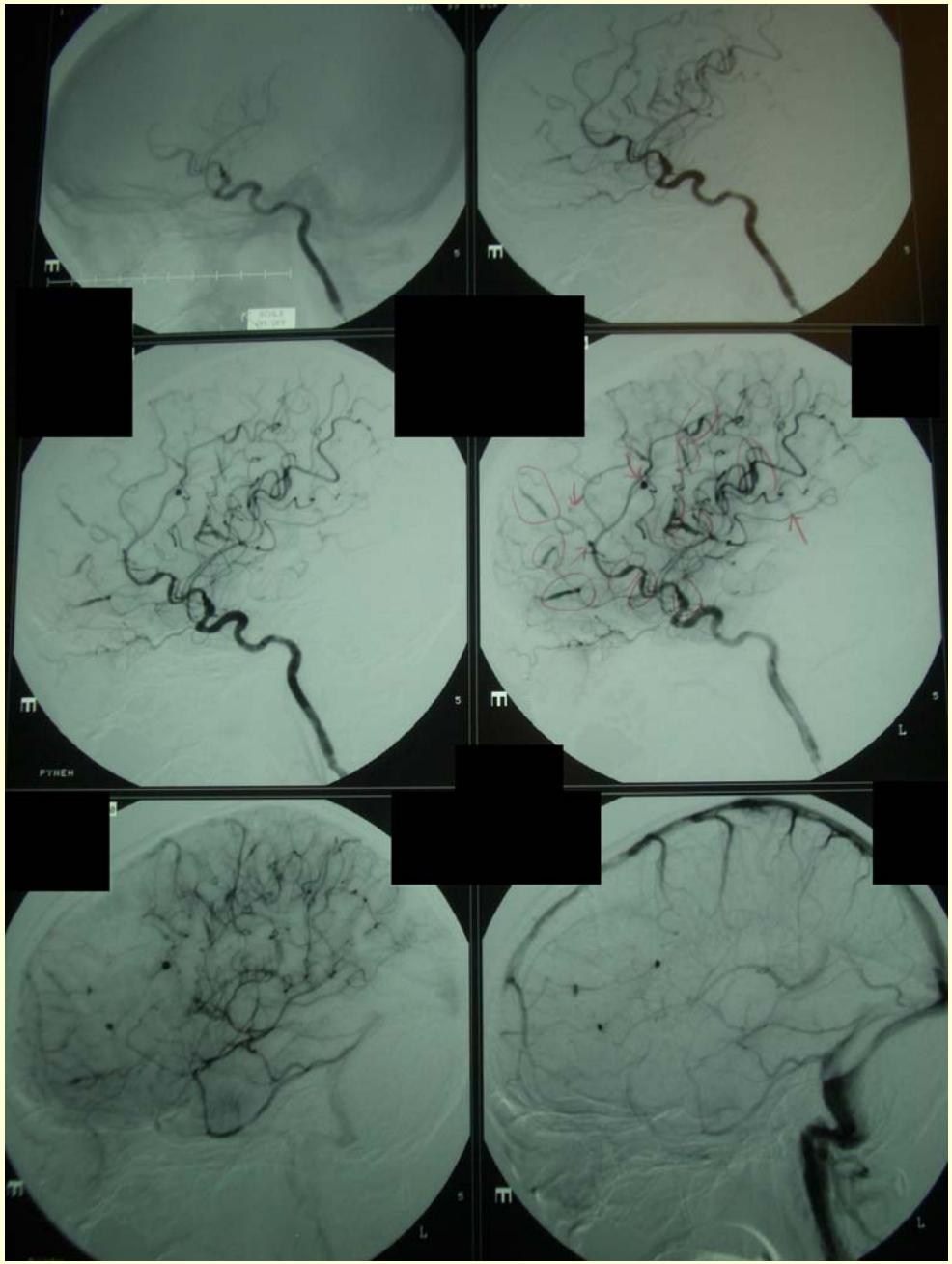
Subacute SAH in left sylvian fissure and medial aspect of left middle cranial fossa

Cause of SAH ??

- Rupture aneurysm
- Cerebral vasculitis



DSA



DSA

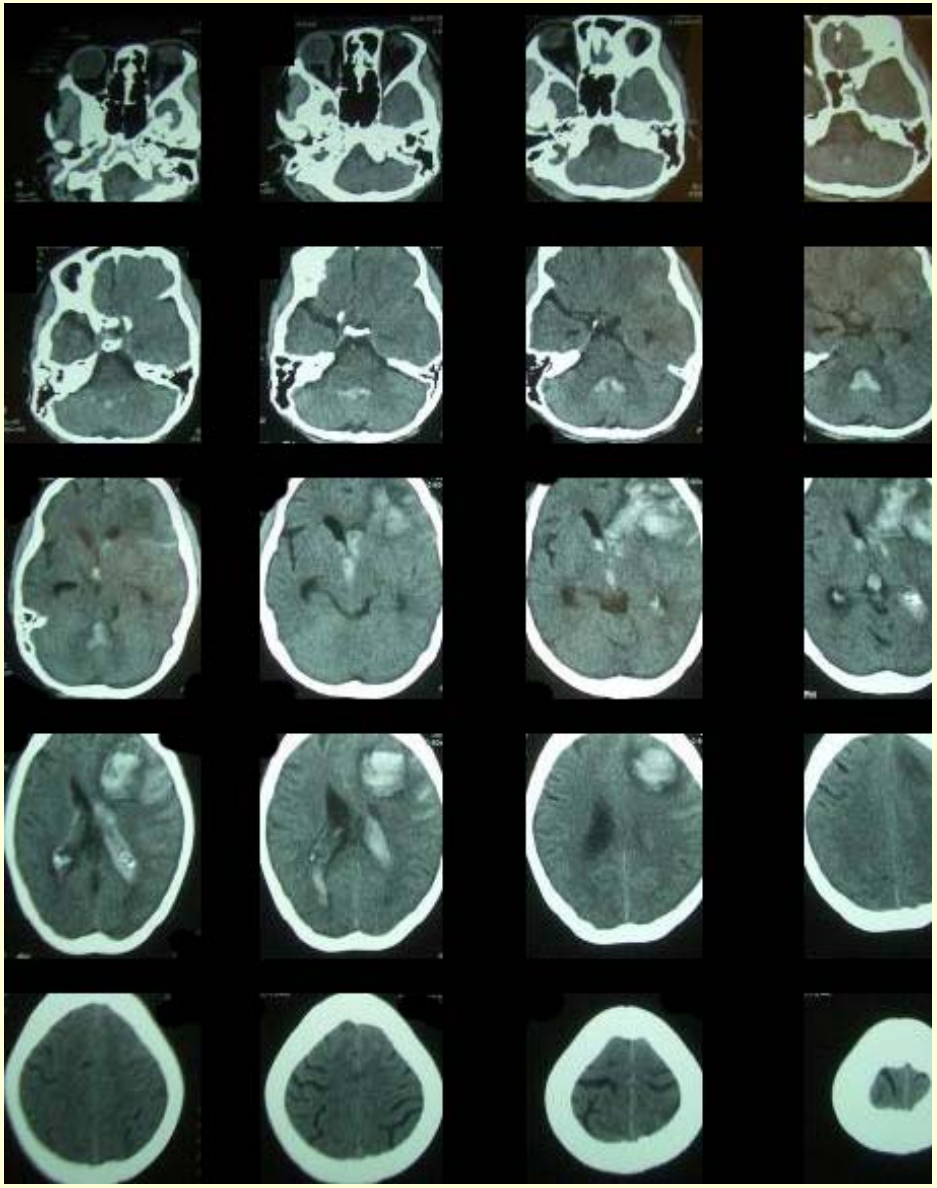
>7 fusiform aneurysms in insular and proximal cortical branches of R MCA
6 L ACA, parietal branch of L callomarginal artery
3 L PCA
2 L MCA

=>The angiographic features are extremely unusual showing multiple intracranial aneurysms and likely related to SLE with vasculits

Tx: conservative

(Day 5)

- Decrease GCS 9/15
- Repeated CT brain



- Neurosurgical Tx :
  - emergency clot evacuation and excision and of left frontal aneurysm
- Medical Tx :
  - pulse methypred 1g daily for 3 days
  - IV cyclophosphomide

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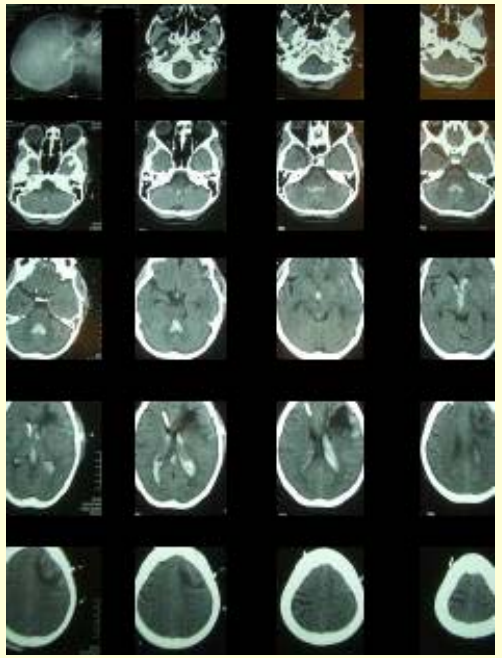
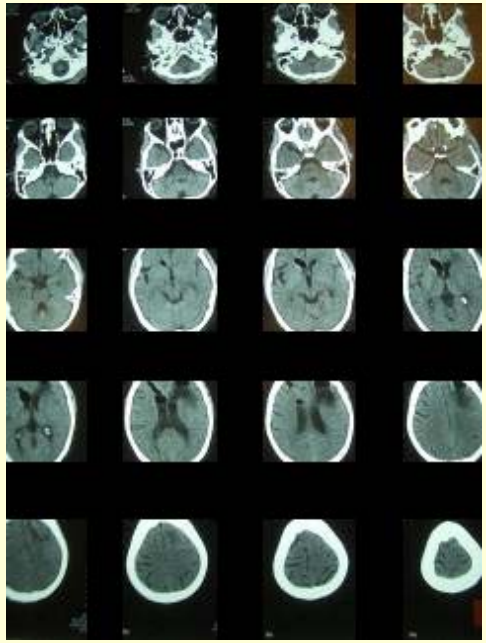
Left frontal artery biopsy

- compatible with rupture aneurysm secondary to lupus vasculitis of intracranial vessels

progress

- Post operatively, gradually regain full consciousness
- Extubated on day 1 post op
- Monthly CTx for 4 more doses given, then switch to monthly IVIg because of low WCC after CTx





- 2009 → ADL indep, work as clerk, no focal neurological deficit

Unanswer question

- Will the 2nd rebleed be prevented ?????

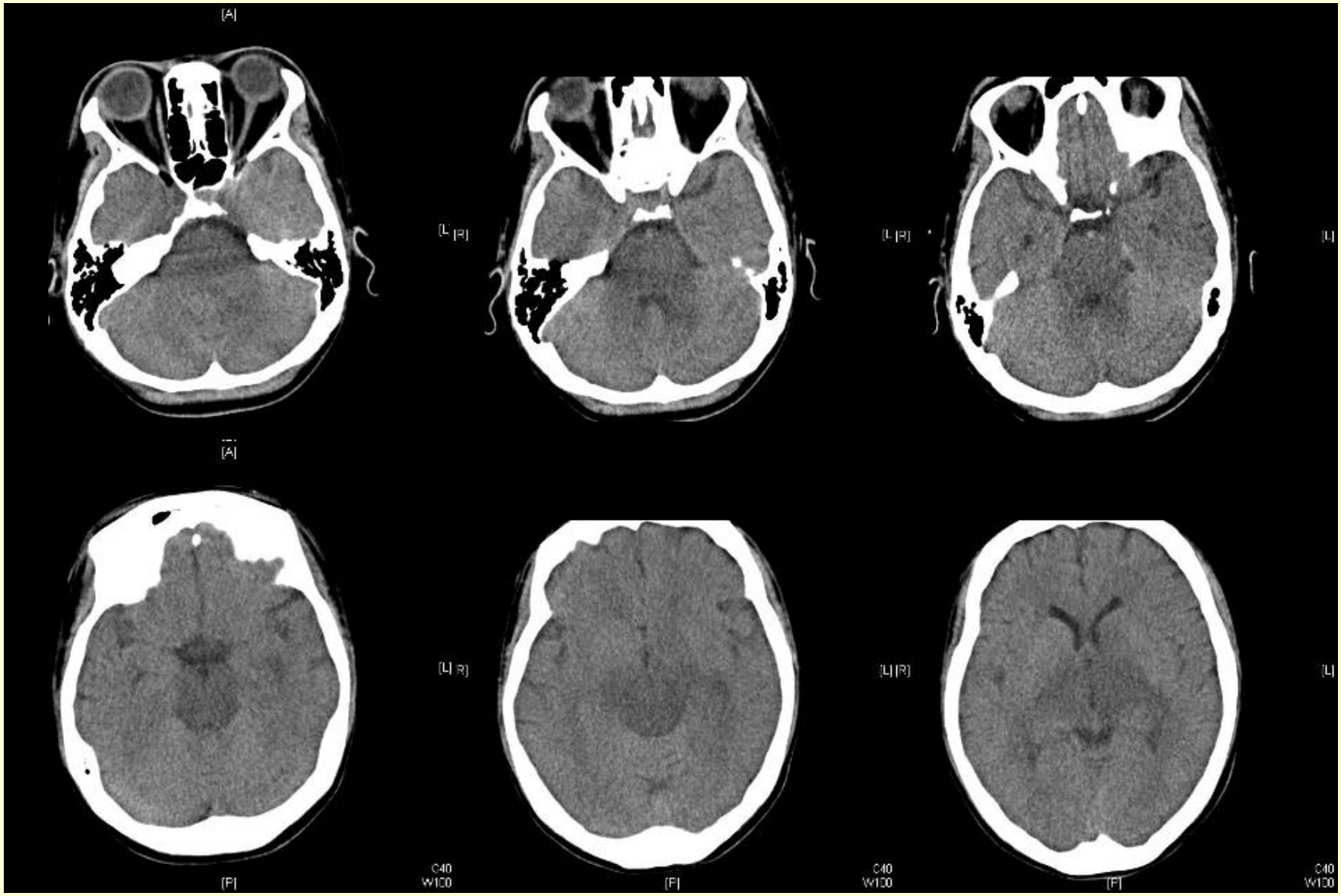
Case 2

- F/25
- SLE with class IV lupus nephritis treated by private rheumatologist with Myclophenonate Mofitel at age of 22
- Relapse of lupus nephritis 2 yrs later, see herbist for 6 months with no improvement, admitted to PWH because of severe edema and was diagnosis to have ESRF 5/2008 required HD. CAPD started.
- HT poor drug compliance

c/o

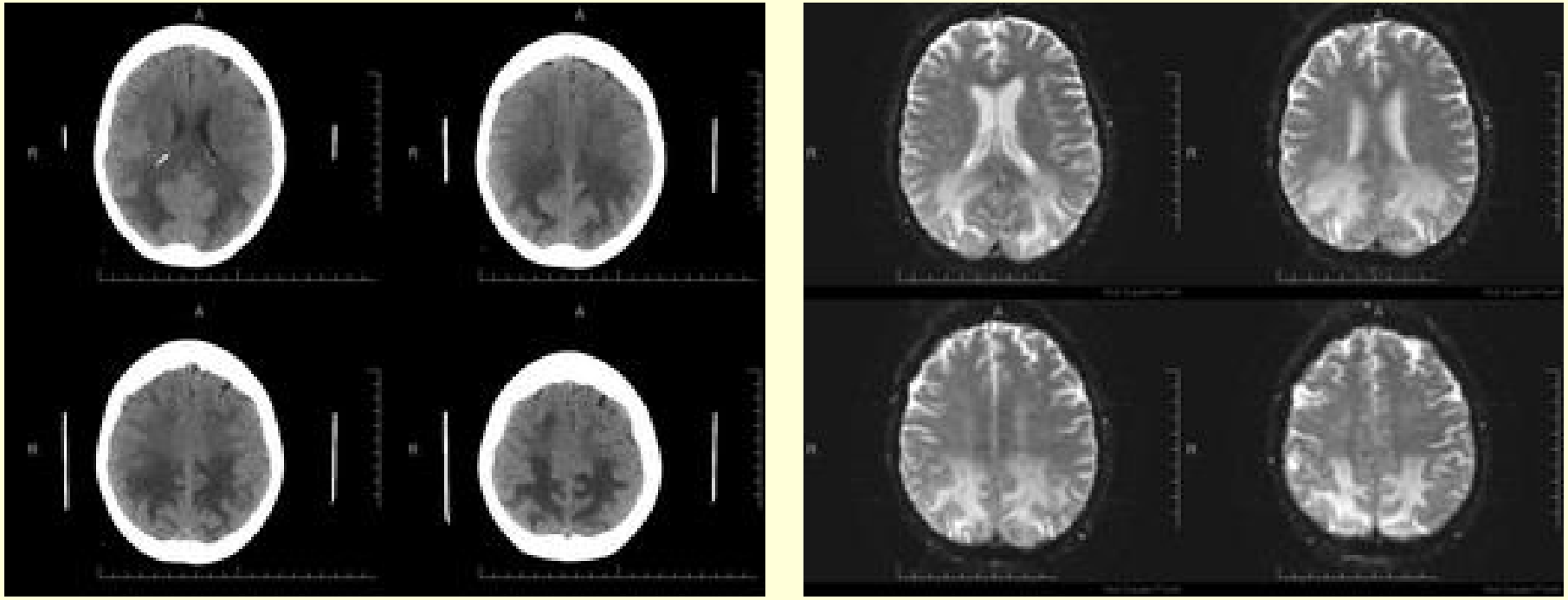
- confusion at home
- Develop convulsion at A&E, abort by valium
- In medical ward, convulsion again with decrease GCS 5/15,
- BP 190/100
- Consult ICU for airway protection

CT brain



- DDx:
  - Cerebral infarction
  - Sinus thrombosis
  - Cerebral vasculitis
  - Posterior reversible encephalopathy syndrome (PRES)

MRI brain



T2-weighted image shows hyperintensity in the white matter of the occipital lobes.

progress

- Control BP
- Regain conscious afterwards

Posterior reversible encephalopathy syndrome (PRES)

Association:

- Uncontrolled hypertension
- Acute or chronic renal failre
- TTP/ HUS
- Eclampsia
- CTD : SLE, PAN, WG …
- Immunosuppressant / chemotherapy
- Cyclosporin A, Methotrexate, Tacrolimus, IVIg

PRES

- Clinical features:
  - Headache
  - Alter consciousness
  - Visual disturbance
  - Seizures
- Investigation:
  - CT brain
  - LP
  - MRI brain
  - EEG
- Treatment:
  - BP control
  - Dilantin for seizures
  - Decrease dosage or withdraw of culprit drugs

Approach to SLE with renal failure

- Poor prognostic factors for SLE:
  - Lupus nephritis usually class IV
  - Refractory to various immunosuppressant
  - Poor compliance
  - Male
  - Asian, African American

Presentation:

- Acute on chronic renal failure
- Acute renal failure
- Uraemic symptom
- Fluid overload
- hyperK

Precipitation factors:

- Nephrotoxic drugs
- Herbs
- UTI
- Overdiuresis
- Uncontrolled lupus nephritis

Investigation:

- MSU → to exclude UTI
- Urine microscopy → dysmorphic RBC, casts
- 24 hour urine for albumin
- Blood test → including DsDNA, C3 level
- Renal ultrasound

Tx:

- Dialysis support

Case 5

- M/46 labour worker
- Known SLE with class IV lupus nephritis FU RH, poor compliance to MMF
- Admitted PY 8/2008 for lower limb edema, serum alb 28
- Diagnosed as Class IV lupus nephritis, probably refractory to MMF (or poor compliance)
- Switch MMF to intravenous monthly cyclophosphamide
- On schedule 4th dose of cyclophosphamide, admitted clinically for anacasa and deteriorating renal function Cr 250 (baseline130), CXR → bilateral pleural effusion
- Pulse methylprednisolone 500 mg given for 3 doses
- In view of the poor respond to cyclophosphamide, rituximab was given

- 3 days after the rituximab, condition further deteriorate with increasing SOB
- CXR → congested and bilateral plerual effusion
- Cr increased to 430
- ICU consulted for support
- HD offer and NIV supported
- Total 11 kg of fluid drawn out through the HD session

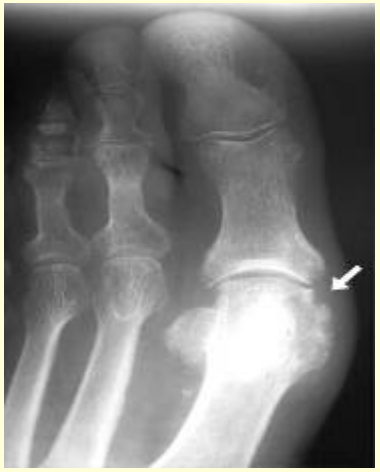
- However, complicated with PTB, further doses of rituximab was not given
- After PTB was treated for 4 weeks
- Rechallenge with low dose MMF + tacrolimus
- Gradual improved after 3 months with serum alb 15 → 35 though the Cr remained 280+

Approach to patient for suspected autoimmune disease

Rheumatoid arthritis (RA)

RA – diagnostic criteria

1. Morning stiffness
2. Arthritis of 3 or more joints
3. Arthritis of hand joints
4. Symmetric arthritis
5. Presence of rheumatoid nodule
6. Positive rheumatoid factor
7. X-ray erosion



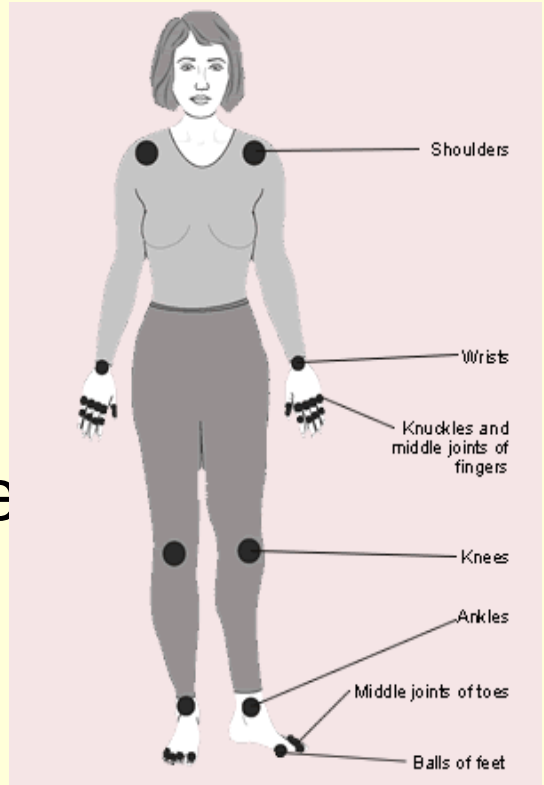
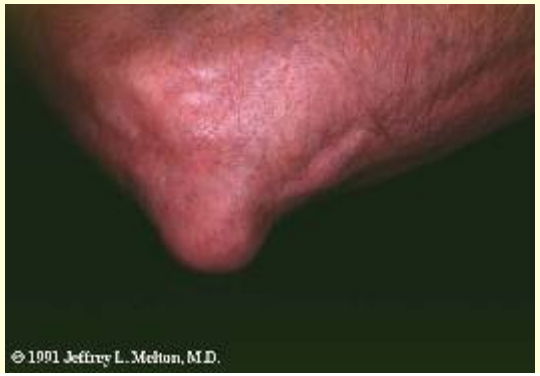


Figure 4. Joint frequently affected by rheumatoid arthritis. Less commonly affected are elbows, hips and the neck.



1987 ACR diagnostic criteria for RA
→required 4 out of the 7 criteria above

RA- treatment

- Pharmacological treatment
  - Pain killer
    - Simple analegesic
    - NSAID
  - DMARDs
    - Hydroxycholoquine
    - Sulphasalazine
    - Methotrexate
    - Leflunomide
    - Cyclosporine
    - Azathioprine
    - Cyclophosphamide
  - Biologics
    - Anti-TNF (infliximab, enbrel, humira)
    - IL6
    - rituximab

Potential life threatening condition in RA patients

- Severe infection
- Intubation
- Respiratory failure
  - Pneumonitis (e.g MTx induced)
    - any time at any dosage
    - nonproductive cough. fever, dyspnea, hypoxemia, or pulmonary infiltrate
  - Interstitial lung disease

Severe Infections in rheumatoid arthritis

Most common infections:

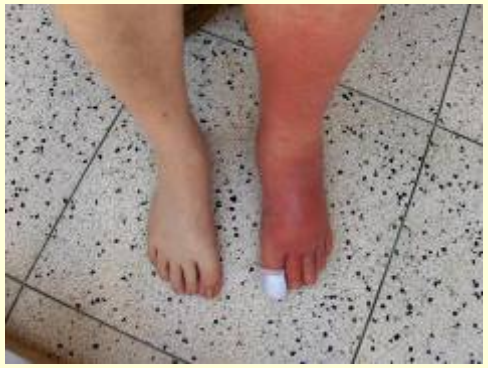
- Pneumonia
- UTI
- Septicaemia
- Cellulitis
- Septic Arthritis

Tx principle:

- Antibiotic – broad spectrum
- Prednisolone – stress dose
- Withold immunosuppressants

  * neutropenic fever* if due to drugs
    - G-CSF if Neutrophile count < 0.5
    - MTx – folinic acid rescue, 15 mg Q6H po
    - Leflunomide – cholestryamine wash out
- Watch out for opportunistic infection esp on biologics

Case 3

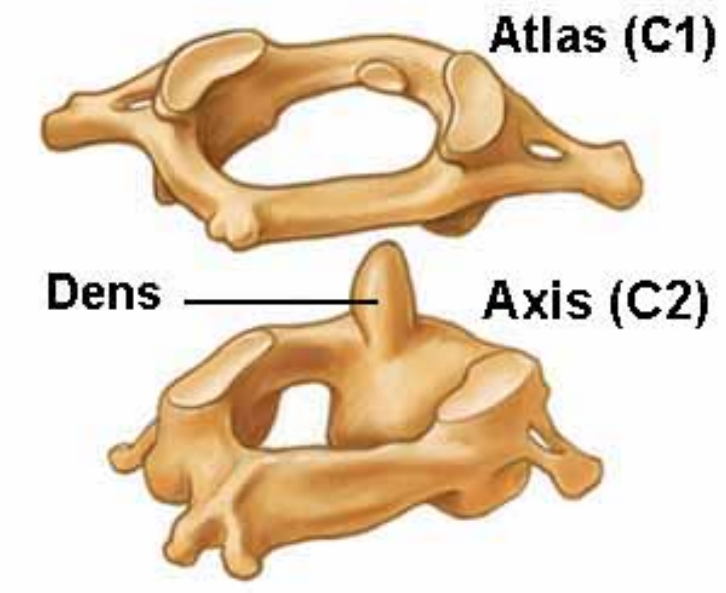


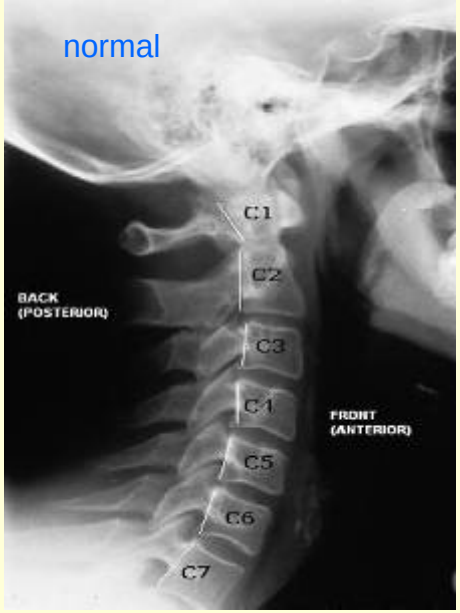
- F/45
- Obesity
- DM on metformin + diamicron
- Rheumatoid arthritis on leflunomide 20 mg daily + Methotrexate 20mg weekly

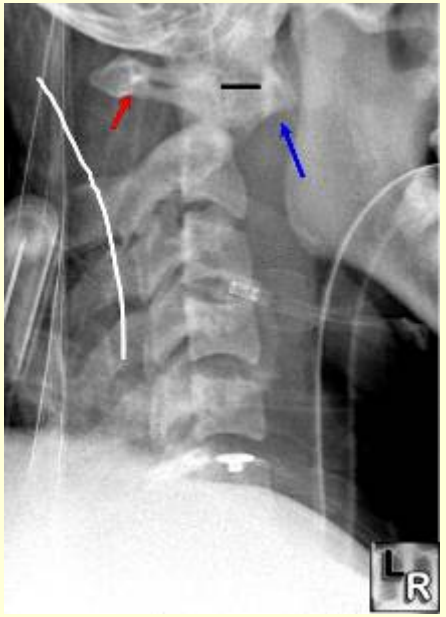
- Admitted for left lower limb cellulitis after toe-nail cutting
- Given IV ampicillin + cloxacillin
- Poor response with extension of the cellulitis up to left thigh, septicaemia shock
- Transfer to ICU for further care
- Still uncontrolled sepsis with ARDS, ARF, VAP and required above knee amputation.
- Post operatively, prolonged stayed in ICU, then was taken over to F6 respiratory bed for weaning of tracheotomy

Intubation in rheumatoid arthritis

- C1-2 subluxation
- Definition: (lateral flexion view) Distance between the anterior surface of the dens and the posterior surface of the tubercle of C1 >3 mm in adults and >5 mm in children



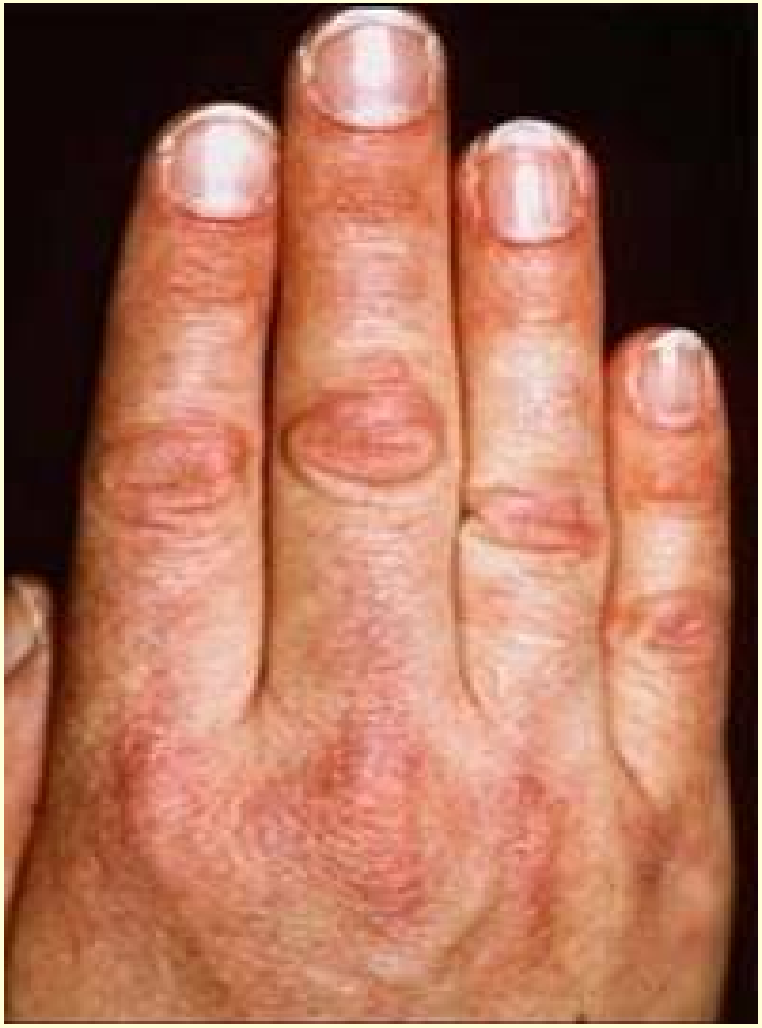


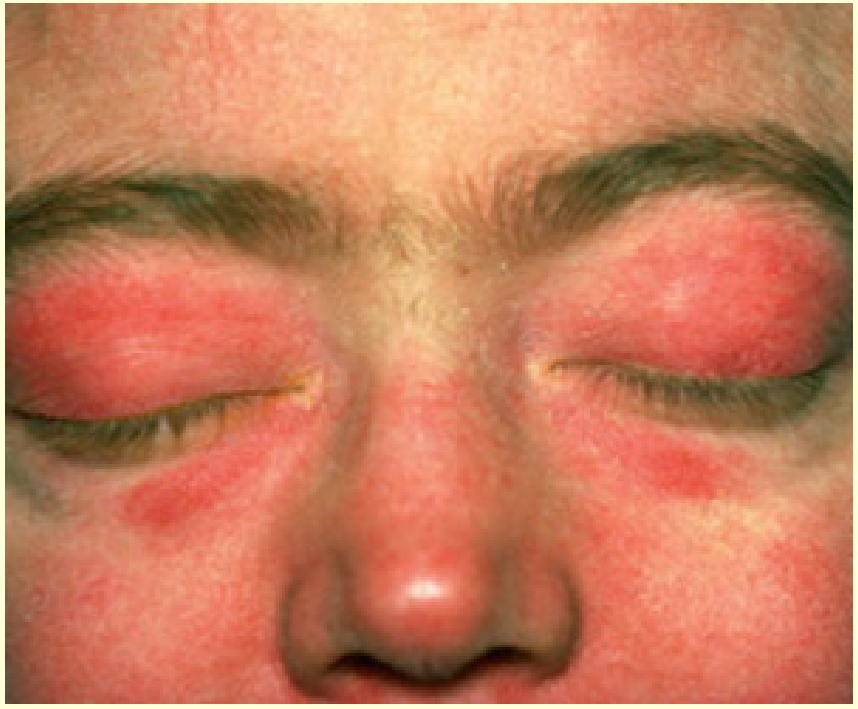


Intubation in rheumatoid arthritis

- Almost all atlantoaxial dislocations involve forward movement of C1 on C2; posterior dislocation is extremely rare
- 21-44%
- 1% neurological impairment
- Risk group:
  - Severe disease :
    - active synovitis,
    - Rapidy progressive erosive peripheral joint disease
    - Early peripheral joint subluxation
    - High inflammatory marker
  - Late onset RA
- Fibro-optic intubation

Dermatomyositis





Clinical features

1) Skin rash
   - Gottron's rash
   - Heliotrope rash
   - Shawl sign and V sign

2) Muscle weakness
   - usually insidious onset
   - typical proximal and symmetrical
   - myalgia / muscle tenderness (25-50%)

3) Lung involvement
   - interstitial lung disease (10%)
   - risk of rapidly progressive pulmonary failure and death

4) Esophageal involvement
   - dysphagia, nasal regurgitation, aspiration → increase risk of aspiration pneumonia

5) Cardiac involvement
   - myocarditis
   - heart failure

6) Other systemic manifestation
   - fever, weight loss, raynand phenomenon, nonerosive inflammatory arthritis

Diagnostic criteria

1. Typical rash
2. Symmetric proximal muscle weakness
3. Elevated CK (normal in 4-6%, correlate with severity of weakness)
4. EMG → classical triad for myositis
    - spontaneous fibrillations
    - Low amplitude, short-duration polyphasic motor potentials
    - Bizarre high freq, repetitive discharges Muscle biopsy
5. Muscle biopsy
    - 20% -ve because of pathy disease
    - Site: affected muscle, usual site quadriceps , detold
    - Biopsying the muscle contralateral to the side having abnormal EMG finding

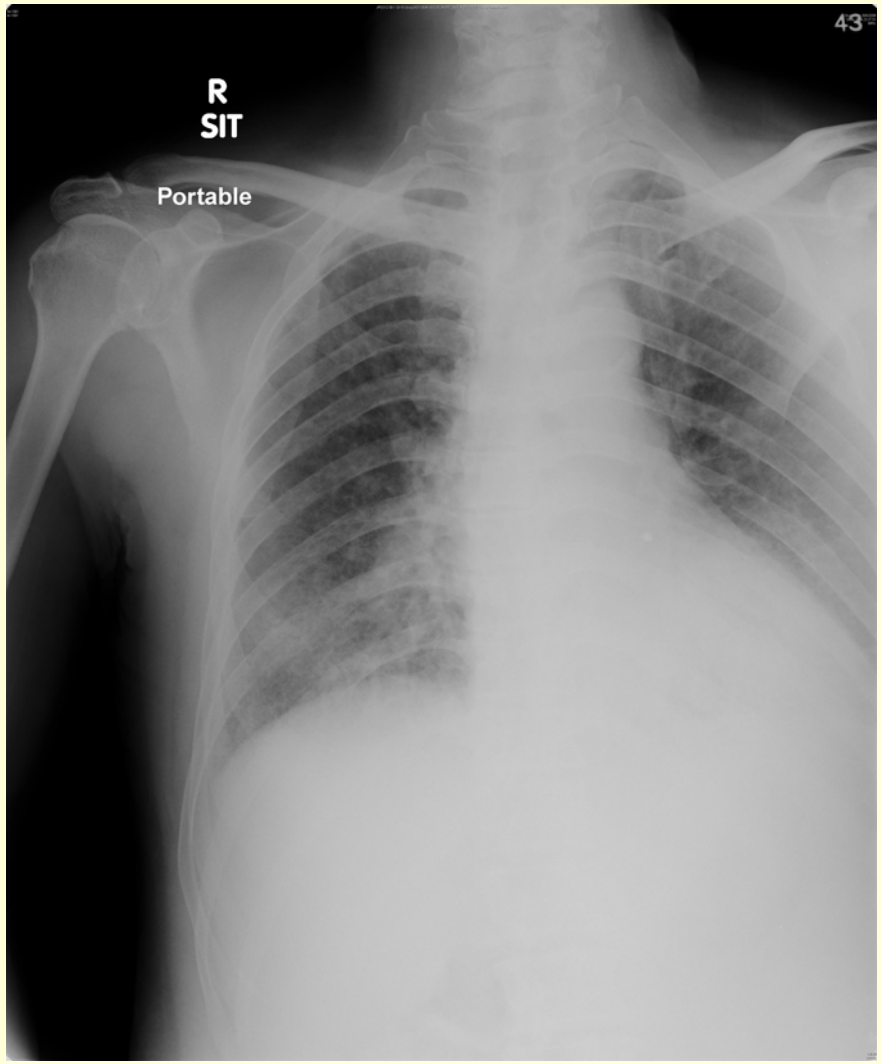
SOB in dermatomyositis

- ? Disease progression
- ? Infection

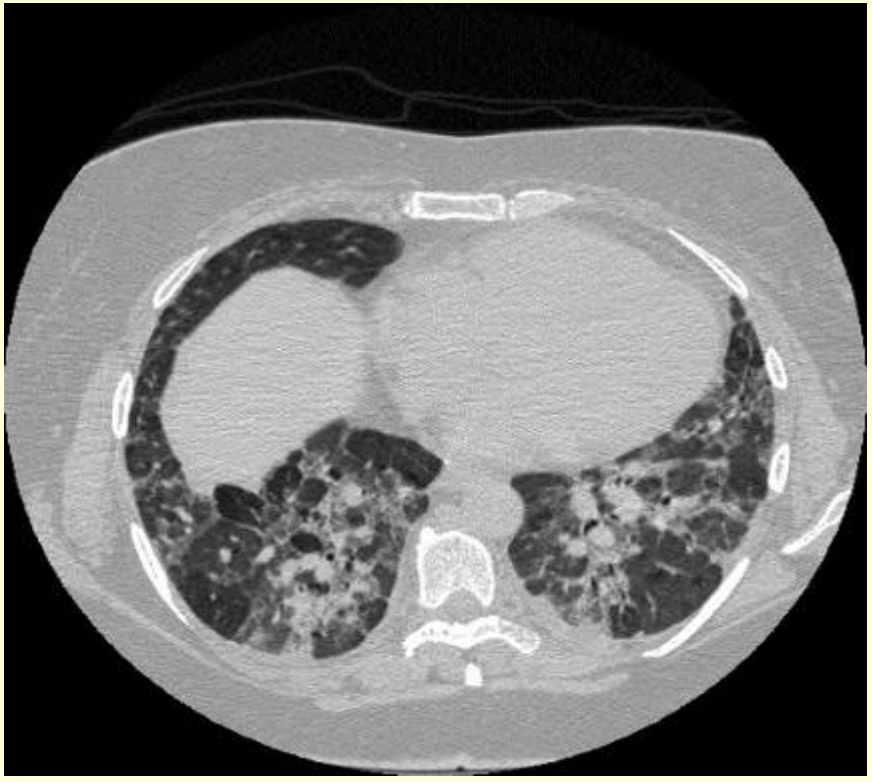
Case 4

- M/60 good past health
- c/o: shortness of breath, generalised malaise and weakness for 2 months
- P/E: multiple raised, scaly, symmetrical skin lesion over knee, back, buttock, neck, hand and elbow
- Chest→ bilateral fine crepitation
- SaO2 88 RA,
- CK 394

CXR on admission



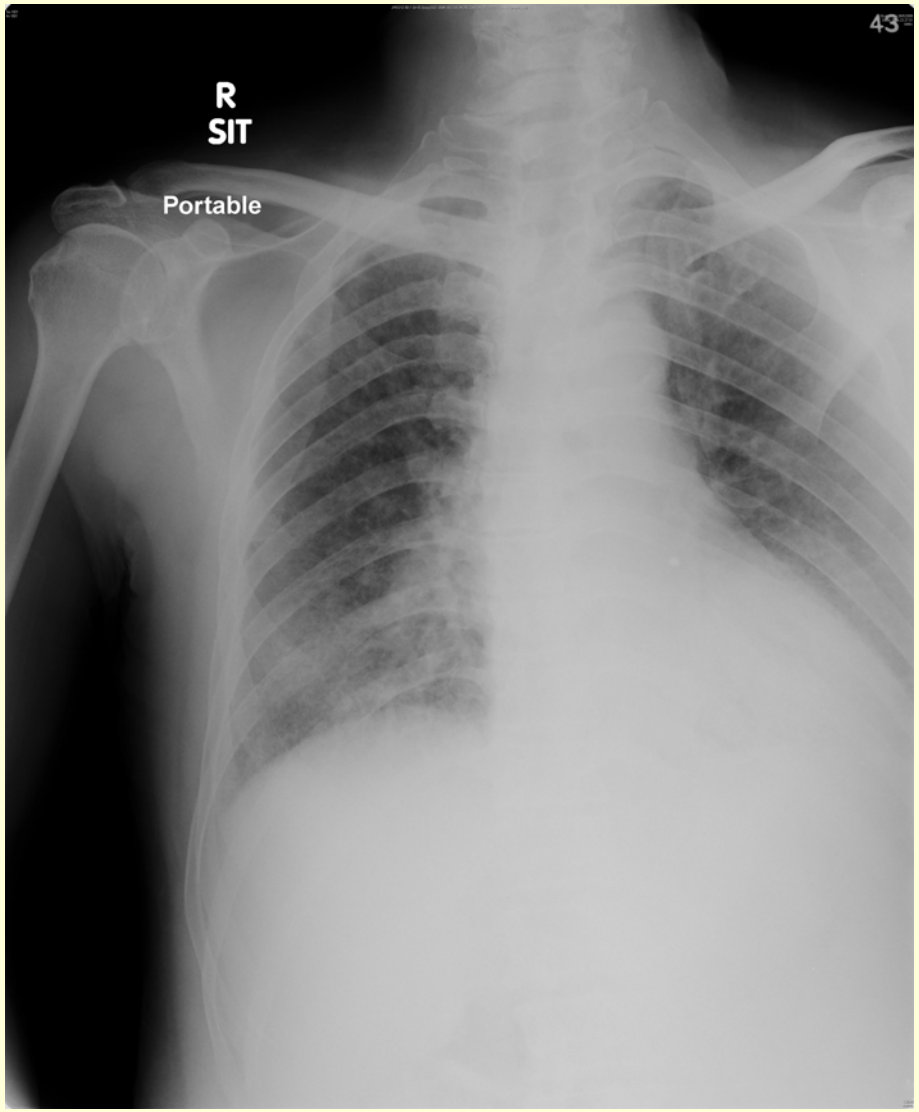
HRCT



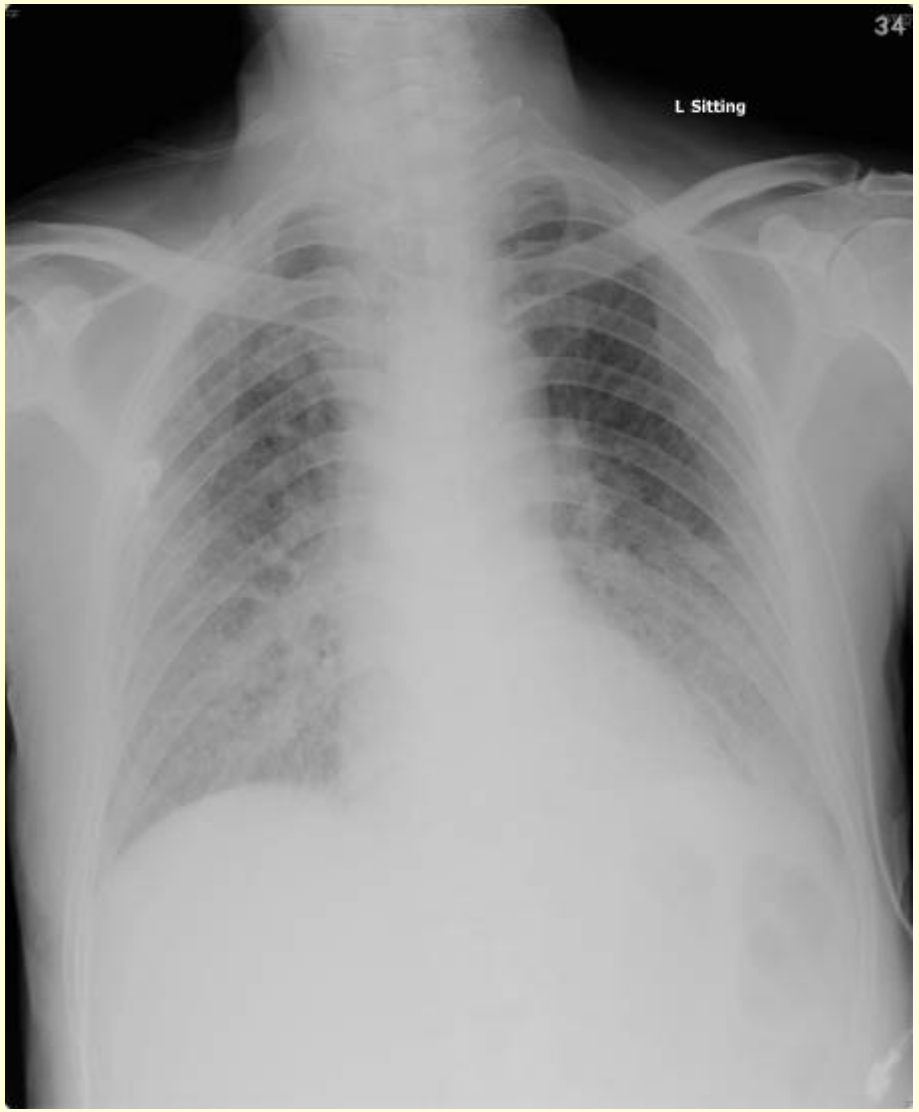
- EMG → evidence of myositis
- Dx: Dermatomyositis with interstitial pneumonitis
- Tx: high dose prednisolone (1mg/kg/day)
- Progress: increasing SOB, desaturation required intubation

CXR

On admission



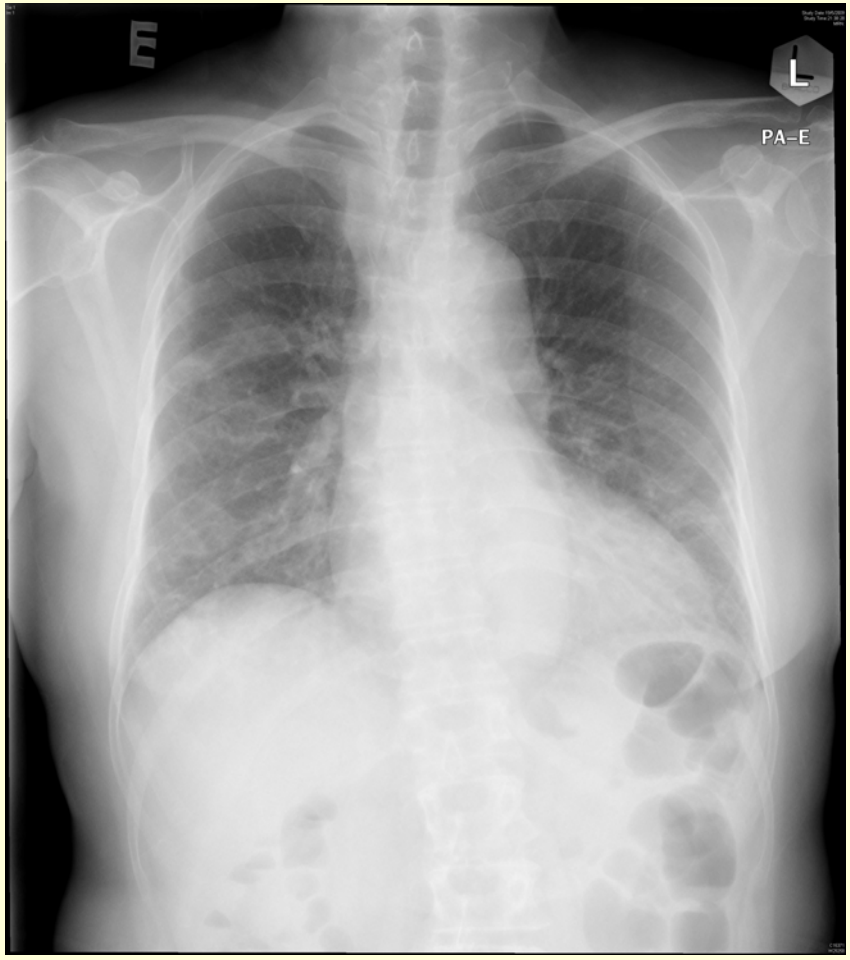
Day 9



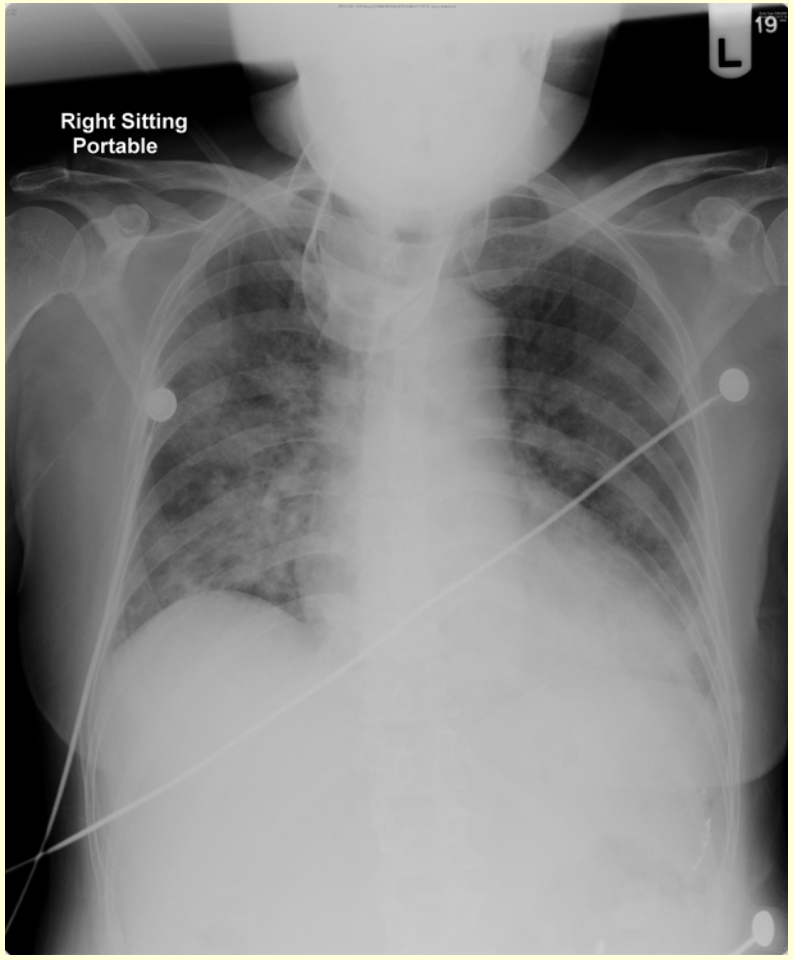
- Impression: rapidly progressing interstitial pneumonitis
- Tx: IV pulse prednisolone 1g daily x 3 days cover with broad spectum antibiotic
- Cx: bilateral pneumo-thorax
- Progress: continue deteriorated and succumbed despite maximum support

Case 5

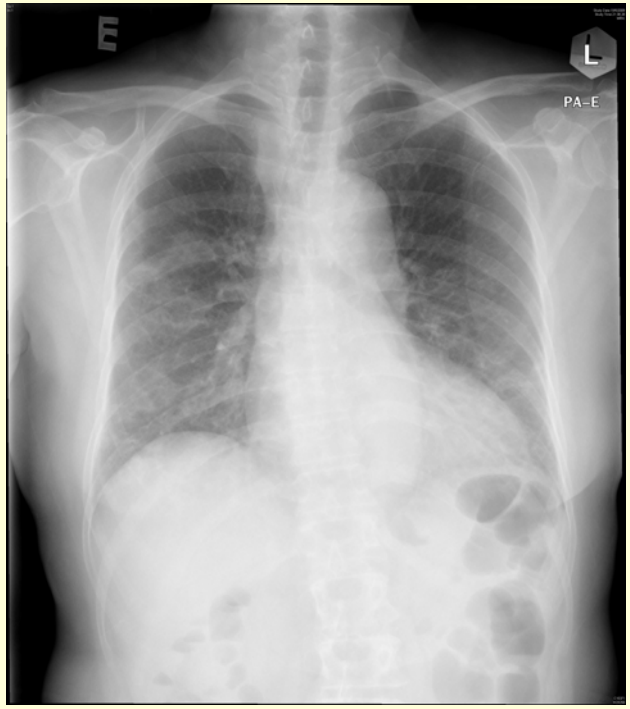
- F/62
- Known dermatomyositis 5/2009 Dx on high dose steroid and Azathioprine



- 1 month later, admitted for fever, cough, sputum and increased SOB



- Treated as chest infection with background Dermatomyositis + ILD
- Required high O2 and contacted ICU for support and required intubation
- Tazocin, azithromycin, tamiflu, amantadine given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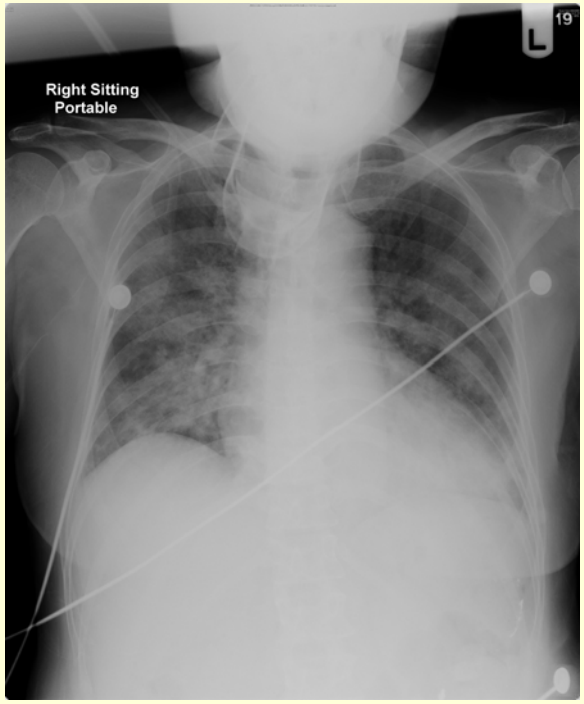
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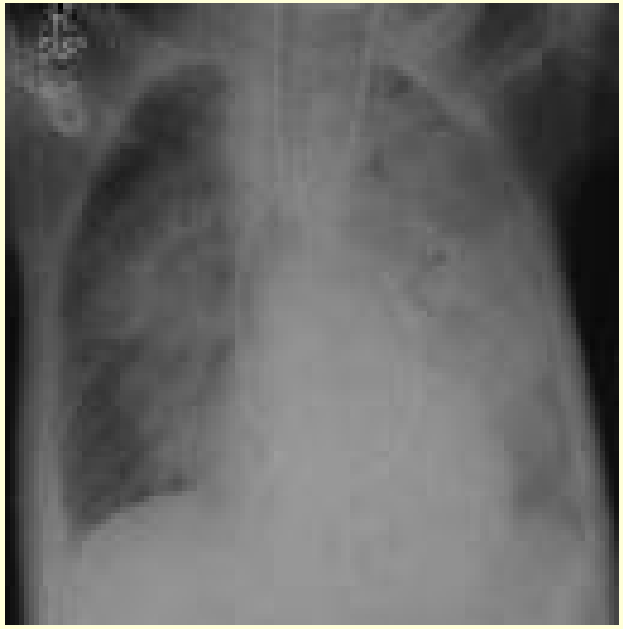
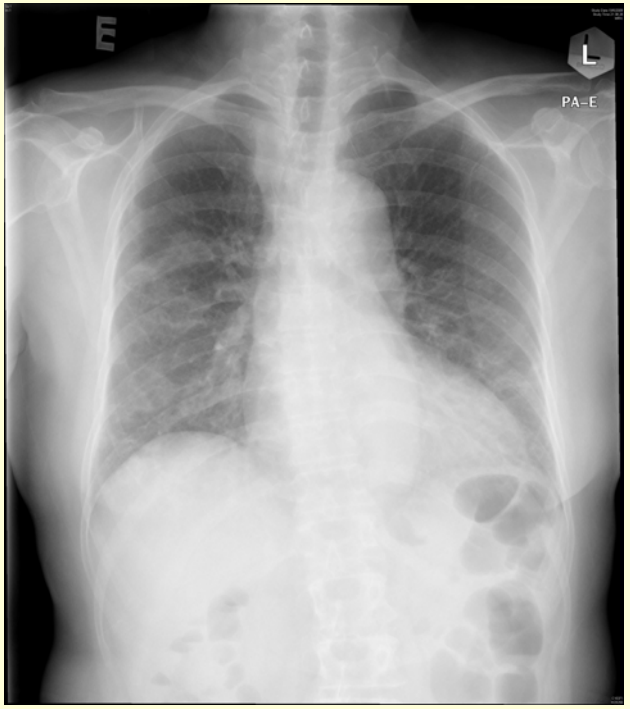
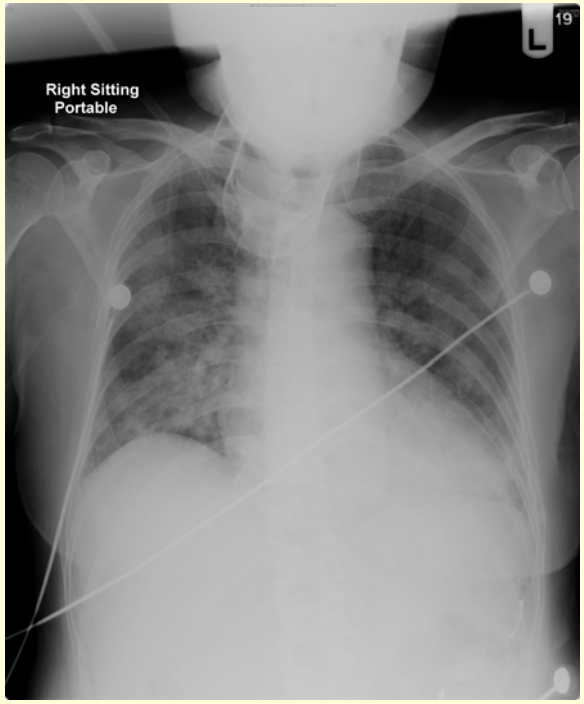
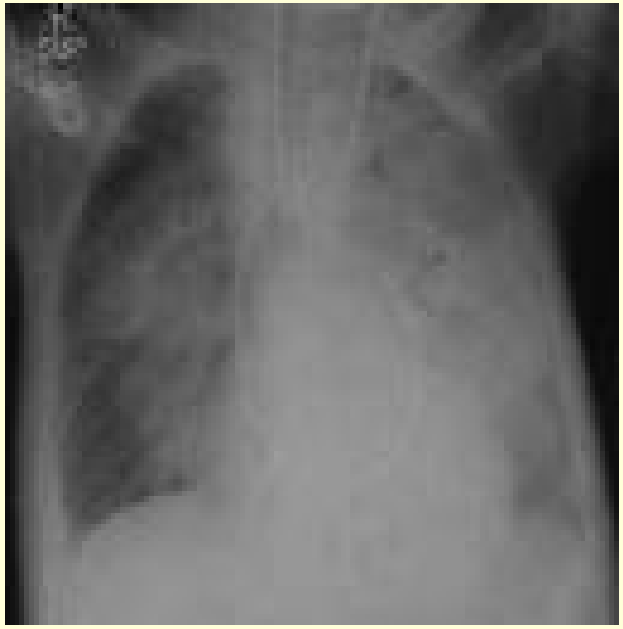








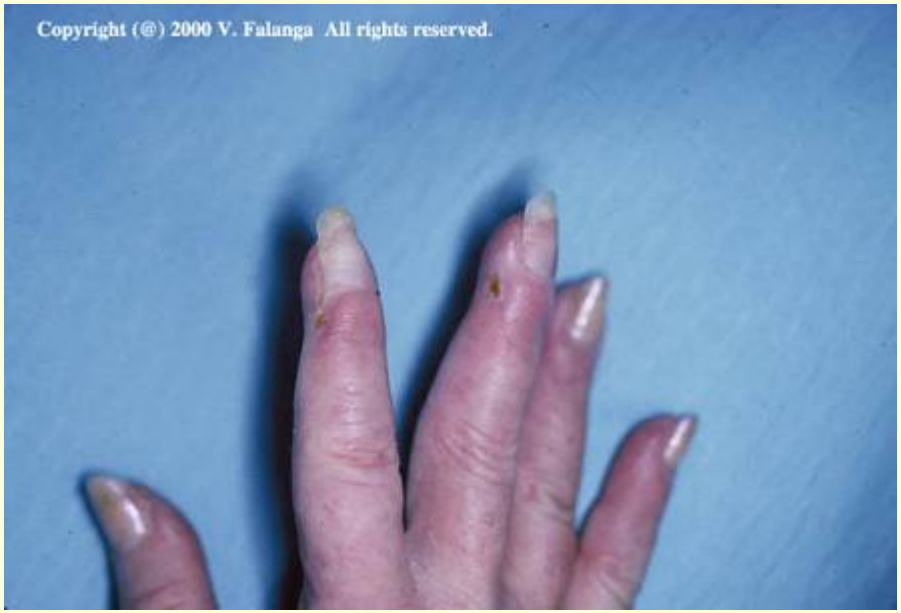
- Continue deteriorate and succumbed

Whenever deteriorate, Had to differentiate flare up of disease vs Infection

High risk infection

- High dose immunosuppressant
- Diseased lung
- Bulbar involvement → aspiration pneumonia

Scleroderma



- Major organ involvement
- Renal → renal crisis
- Lung → interstitial pneumonitis, pulmonary fibrosis
- Cardiac → pulmonary hypertension

Case 6

- F/49 good past health
- Admitted orthopaedic ward for digital ulcer
- Consulted rheuamtologist for raynand Phenomenon
- C/F: sclerodactyly, tighten skin over face, chest wall
- Dx: scleroderma
- Started on aspirin, norvasc

- Admitted for SOB, lower limb edema
- Self detected high blood pressure at home 180/110
- P/E dyspnoea, LL edeam, chest → bilateral creps
- CXR → congested
- Hb 7+
- Cr 1000

- IMP: scleroderma related renal crisis
- Consulted ICU for renal support
- HD and ACEI started
- However, poor response and succumbed

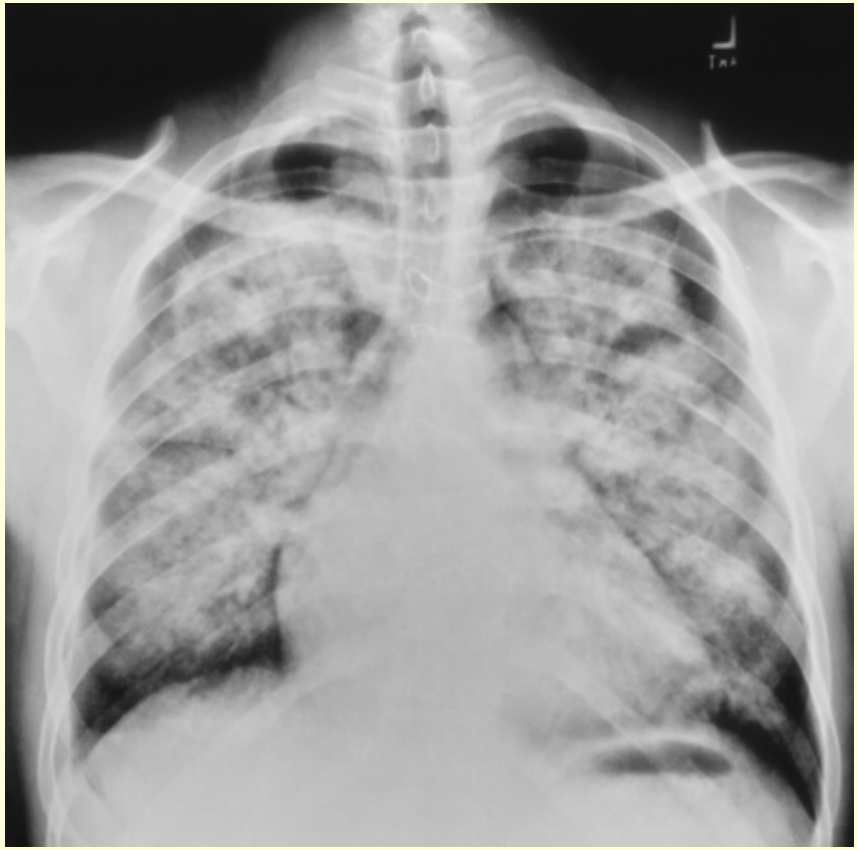
- Risk factors for renal crisis
  - Diffuse skin involvment
  - High dose prednisolone use (>20mg/day) in preceding 6 months
  - Cyclosporin use

Life threatening rheumatic disease diagnosed in ICU

- 20% rheumatic disease first Dx in ICU
- Rheumatic disease sometimes is life threatening
- High index of suspicious is required
- How to interpret the investigation result

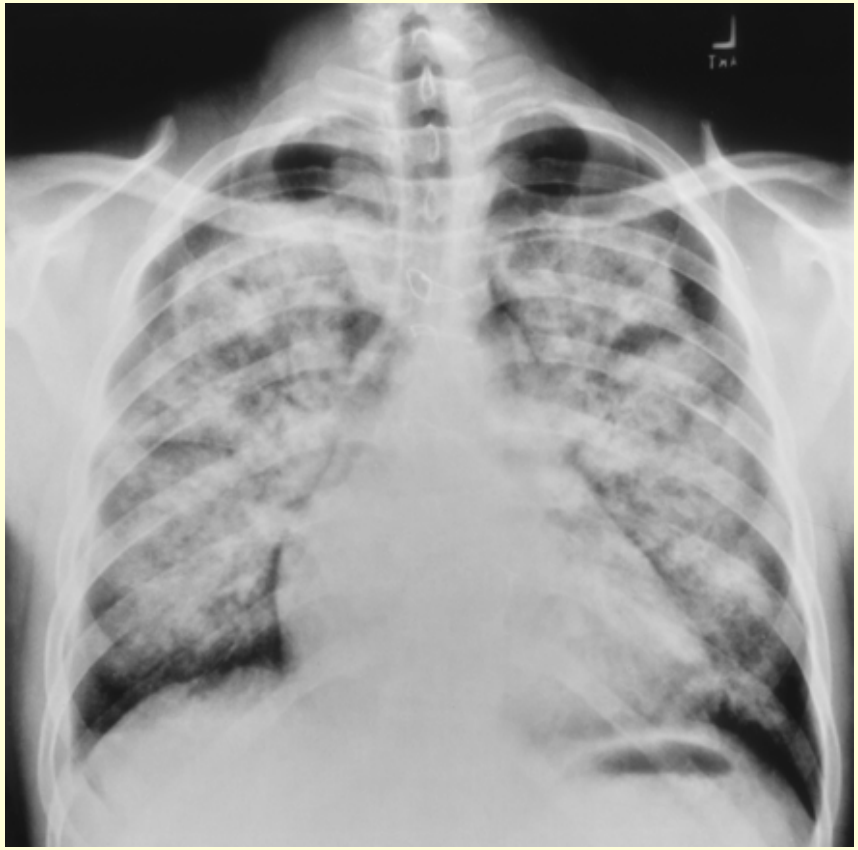
Case 7

- F/61
- Good past health
- c/o SOB, cough, fever , haemptysis
- Hb 7.2



Case 7

- F/61
- Good past health
- c/o SOB, cough, fever , haemptysis
- Hb 7.2
- Urine → RBC +++
- Bronchoscopy → capillaritis, bronchiolitis and organising pneumonitis
- BAL → hemosiderin-laden macrophages
- Urgent cANCA +ve
- Dx: Wegener's granulomatosis



Treatment:

- pulse methylprednisolone
- Oral cyclophosphamide
- Dramatic improvement

Case 8

- F/26
- Admitted A&E for severe abdominal pain
- Already gasing in A&E required intubation
- PEA (pulseless electrical activity) after intubation
- P/E : distended abdomin with guarding
- Ix: severe metabolic acidosis, AXR → dilated bowels
- Emergency → laparotomy showed small bowel strangulation, adhesion and intestinal obstruction, adhesionalysis and ileostomy done
- Post operative condition remined critical
- Succumbed 10 hrs after admission

- Postmortum diagnosis → Takayasu vasculitis
- Recall : fever and wt loss on and off for 2 months

Common problem don’t forget !!

Common problem don’t forget !!


Thanks!