Rheumatologic emergencies in ICU

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Cases Presentation

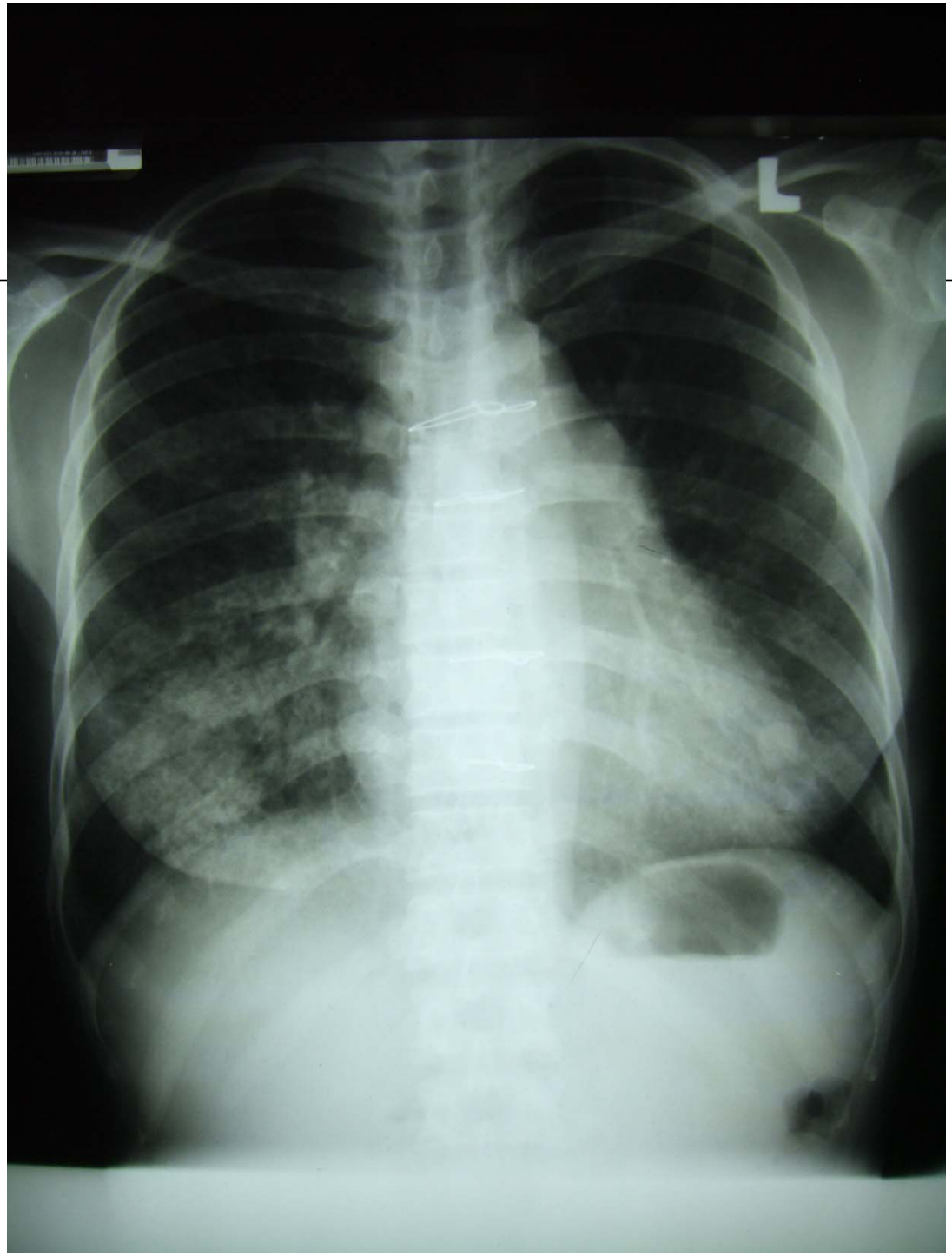
- SLE
- Polymyositis
- Systemic sclerosis
- Antiphospholipid syndrome

Case 1

- Female 40-year-old
- Epilepsy, CRHD with MVR done in mainland China
- On warfarin and Epilim
- Fu in Cardiac clinic for 6 months

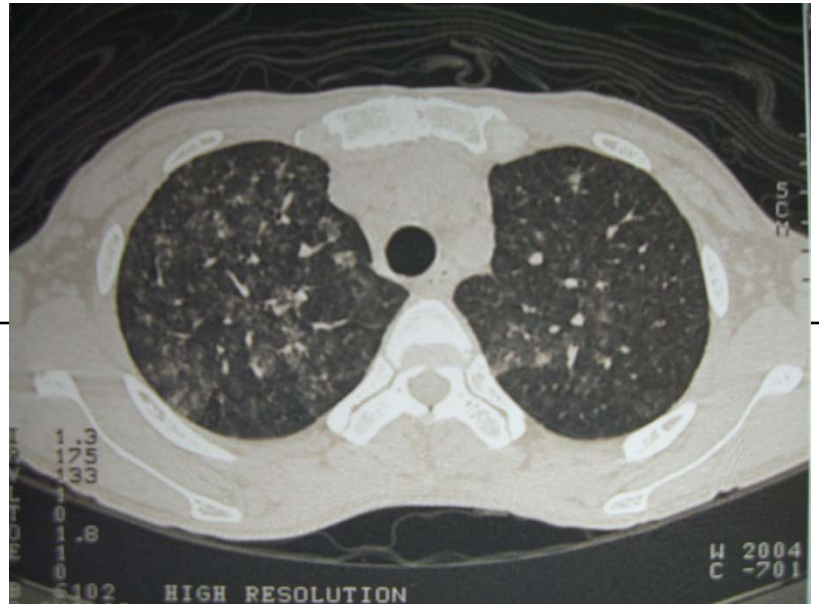
- In Aug 98
- Recurrent episodes of haemoptysis and dyspnea for 10 days
- No fever
- No chest pain

- Hb 6.1, MCV 101, retic 9.6%
- (previous Hb 11.1)
- WCC 2.6, plt 95
- ESR 36
- INR 2.4, APTT normal
- Normal RFT
- Albumin 24
- Urine –ve protein



- Sputum c/st & AFB smear -ve

- Lung function:
  - FEV1/FVC 77%
  - DLCO 89% of predicted
  - DL/VA 138% of predicted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W 2004
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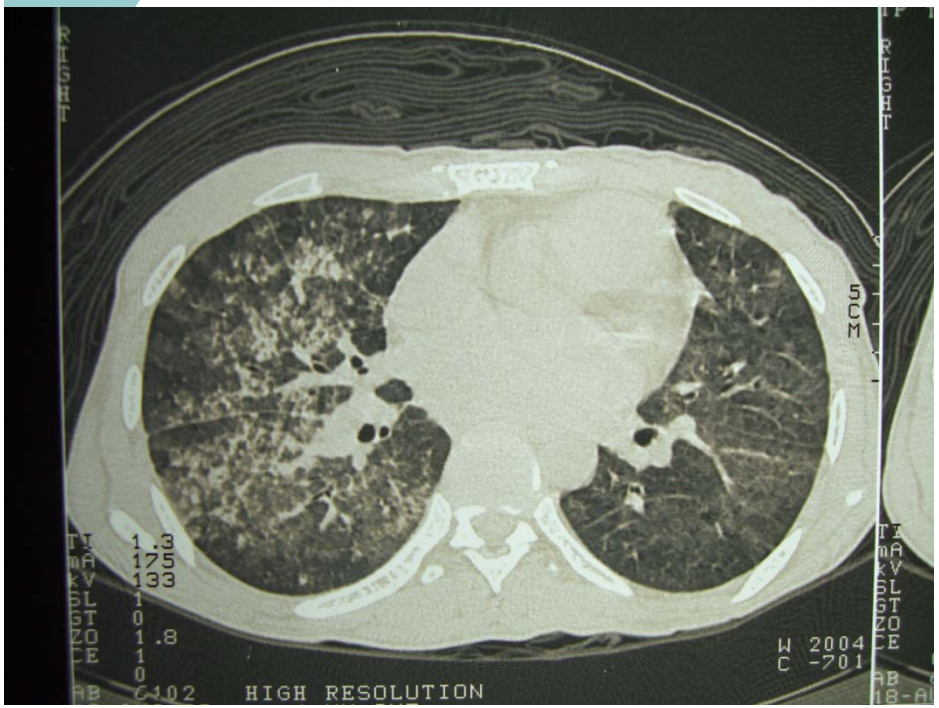
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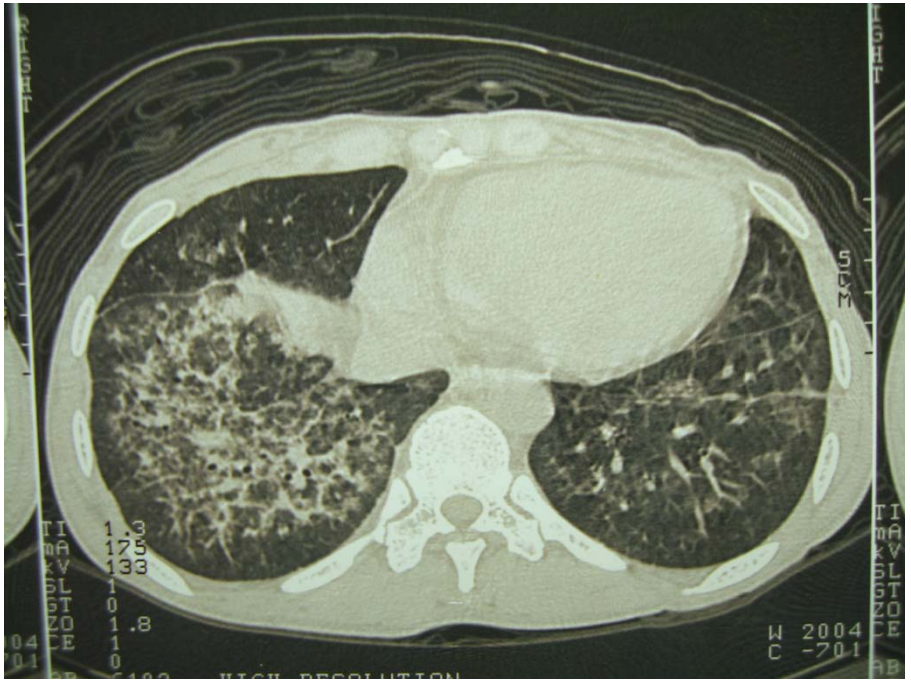
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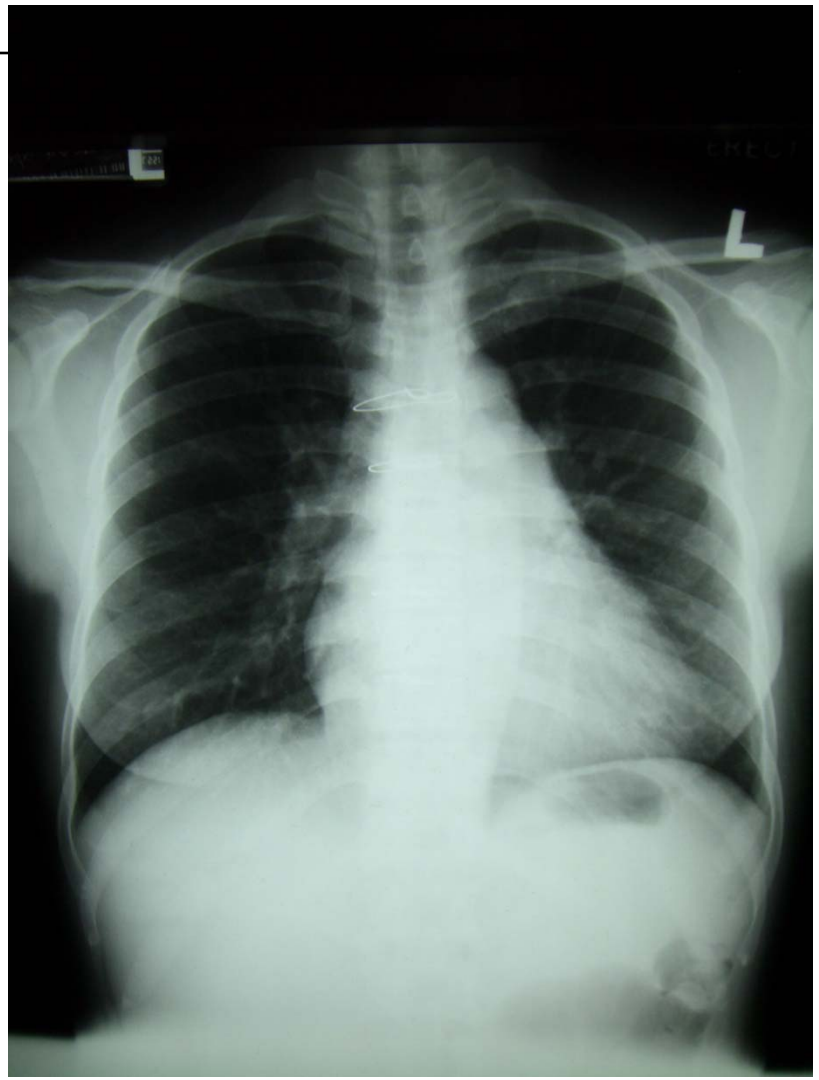
- ANF 1:640, dsDNA 1:160
- Anti-ENA: Ro+
- ↓ C3: 24
- Anticardiolipin Ab –ve
- Anti-GBM -ve, ANCA –ve

- **Bronchoscopy**
  - Scanty amount of sputum, no blood
- **Transbronchial bx**
  - Tiny pieces of lung tissue
  - Alveoli lined by plump pneumocytes, increased fibrosis and siderophages
  - No evidence of infection, vasculitis or malignancy

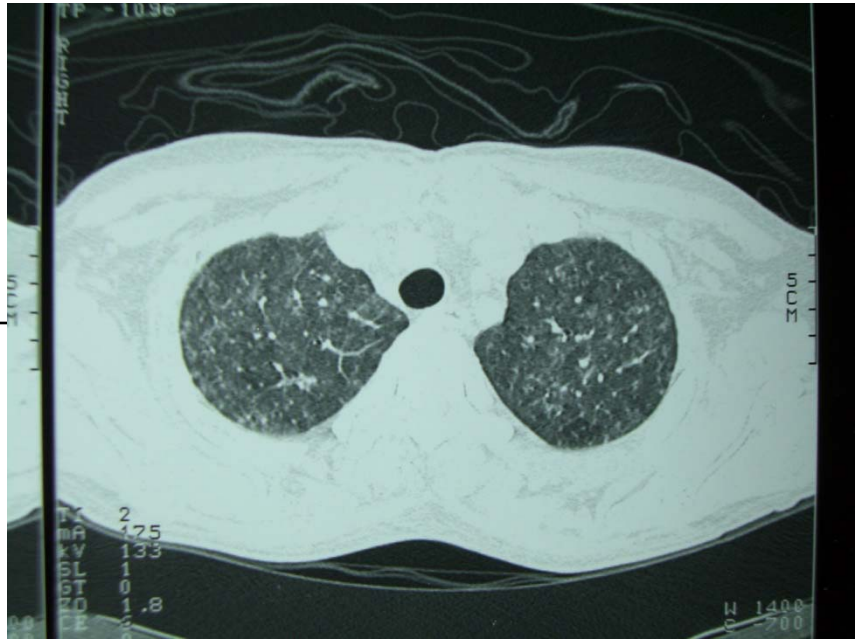
- **SLE**
  - Pancytopenia, abrupt drop in Hb
  - Pulmonary haemorrhage

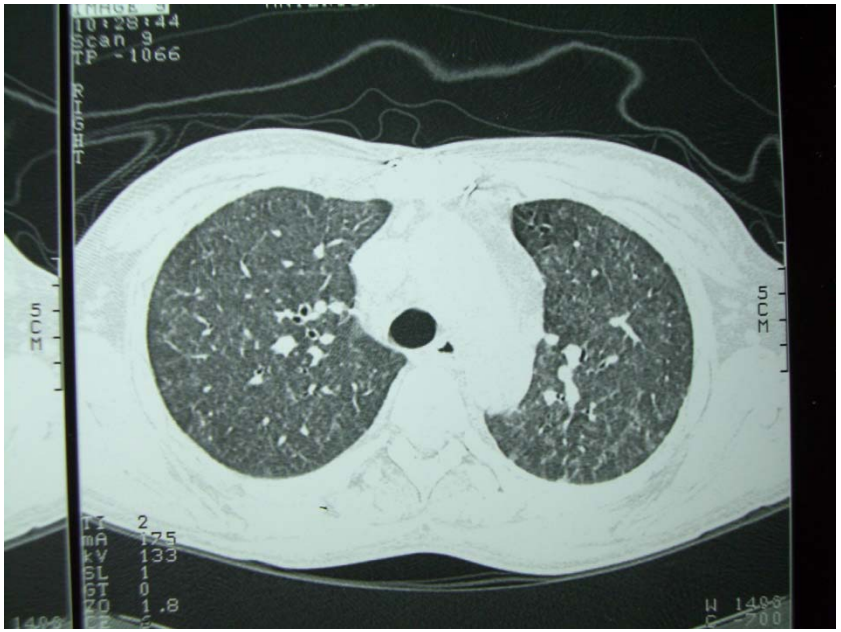
- Stopped warfarin
- O_2 supplement, blood transfusion, antibiotics
- IV Methylprednisolone for 500mg daily for 3 doses
- Prednisolone 40mg daily

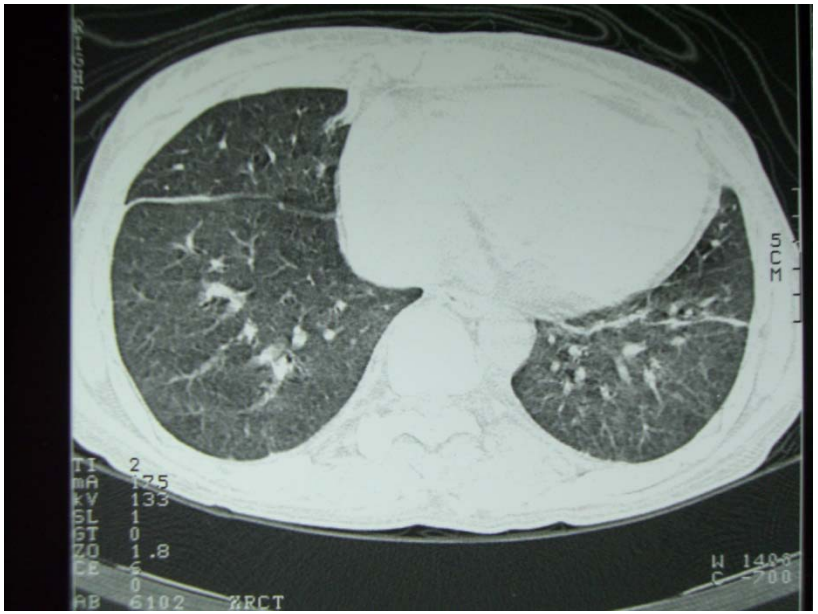
- CXR after Rx



- CT after Rx







Pulmonary manifestations of SLE

- **Pleural**
  - Pleuritis
  - Effusion
- **Parenchymal**
  - Acute lupus pneumonitis
  - Pulmonary haemorrhage
  - Chronic interstitial lung disease
  - Lymphocytic interstitial pneumonitis
- **Airways**
  - Bronchiectasis
  - BOOP
  - Obliterative bronchiolitis
- **Vascular**
  - Thromboembolic disease
  - Pulmonary hypertension
- **Secondary**
  - Infection
  - Atelectasis/ respiratory muscle dysfunction/ shrinking lung syndrome
  - Related to cardiac or renal failure
  - Drug toxicity

- A G Rockall, et al. Postgrad Med J 2001;77:621-638

Pulmonary manifestations of SLE - parenchymal

- Acute Lupus Pneumonitis
- **Pulmonary Haemorrhage**
- Chronic Interstitial Lung Disease
- Lymphocytic interstitial pneumonitis

Acute lupus pneumonitis

- Prevalence
  - 1970s:
    - **4%** (2 in 50 patients)
    - **9.3%** (14 in 150 patients)
    - Estes D, Christian CL. Medicine 1971, 50:85-95
    - Grigor R, et al. Ann Rheum Dis 1978, 37:121-8
  - 2000s:
    - **2%** (2 in 120 patients)
    - **8%** (11 in 137 patients)
    - Mochizuki T, Aotsuka S, Satoh T. Respir Med 1999, 93:95-101
    - Kim JS, et al. J Comput Assist Tomogr 2000, 24 (1);9-18

Pulmonary hemorrhage

- A potentially catastrophic complication of SLE with mortality rate exceeding 50%
- Association with autoimmune diseases
  - SLE
  - Goodpasture's syndrome
  - Microscopic polyangiitis ……
- Shares similar clinical, radiological and histolopathological features with acute lupus pneumonitis
- A part of spectrum of lung disease in SLE results from acute injury to the alveolar-capillary unit

Pulmonary hemorrhage in SLE

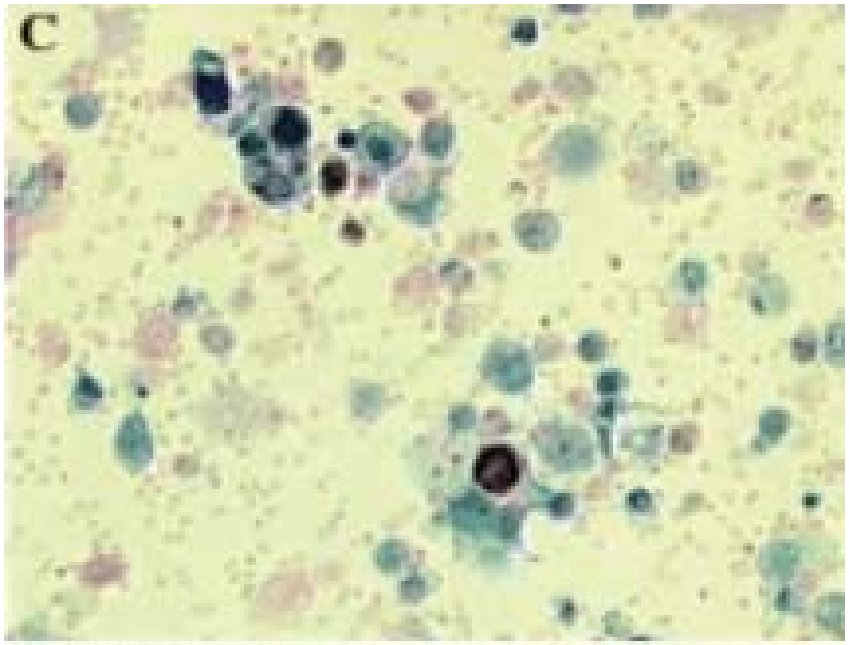
- Rare
- < 2% to 5.4% in cohorts of lupus patients
- Frequently fatal, reported mortality rate 23 to 92%

- Murin S, et al. Clin chest Med 1998; 19:641-665
- Raj R, et al. Crit care Clin 2002; 18:781-803
- Ariger M, et al. Lupus 2004; 13:344-347

Pulmonary hemorrhage

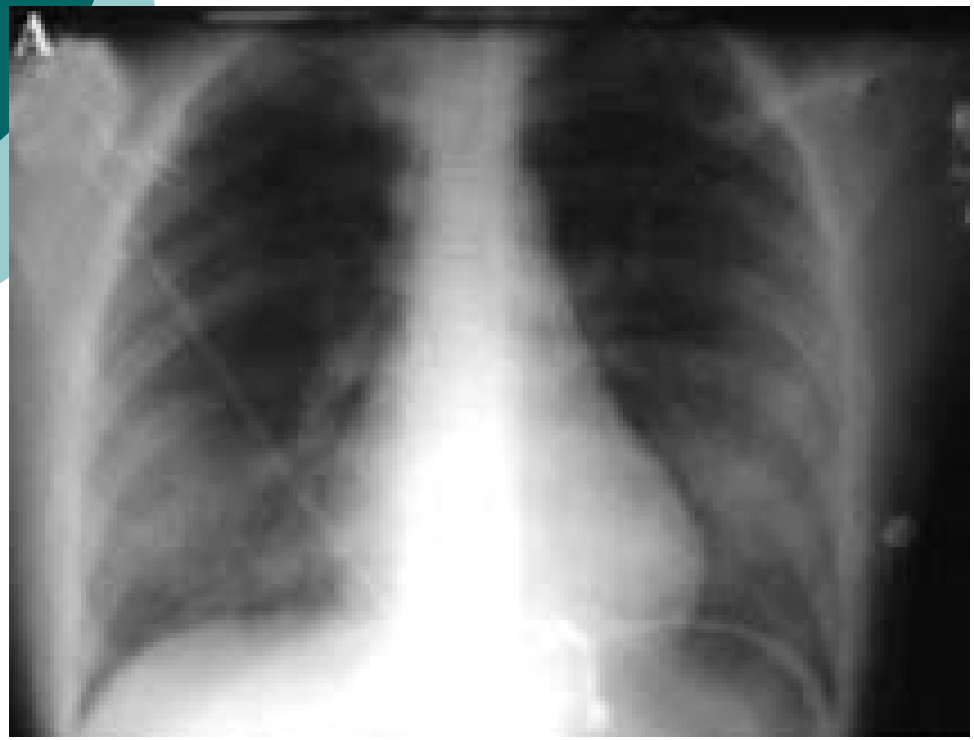
- Hemoptysis in < 50% of cases
- Marked drop in Hb over 12-36 h
- Unexplained rise in DLCO
- Bronchoscopy
  - Blood-filled airways
  - Hemosiderin-laden marcophages in BAL fluid or within alveolar spaces of lung bx specimens

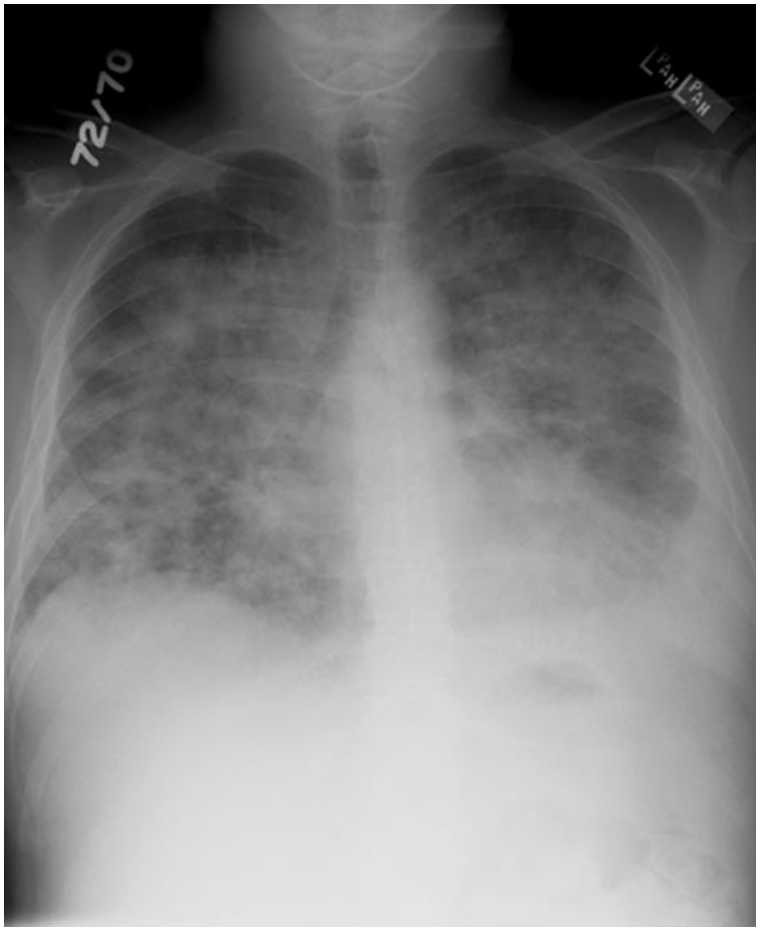
Pulmonary hemorrhage



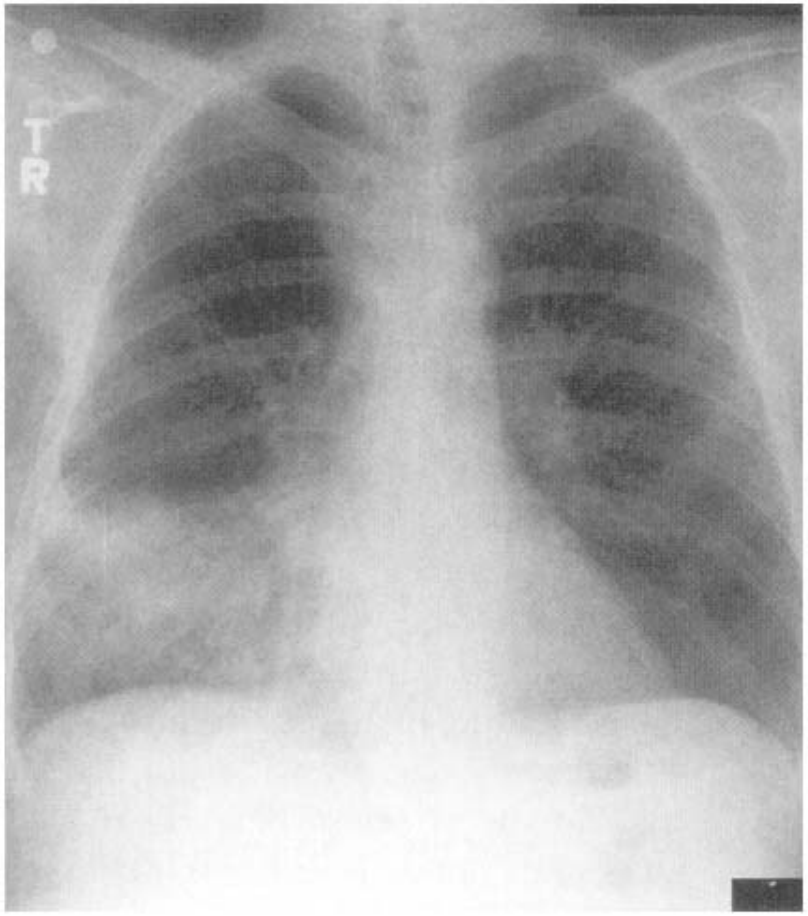
Bronchoalveolar lavage showed red cells with **siderophage**

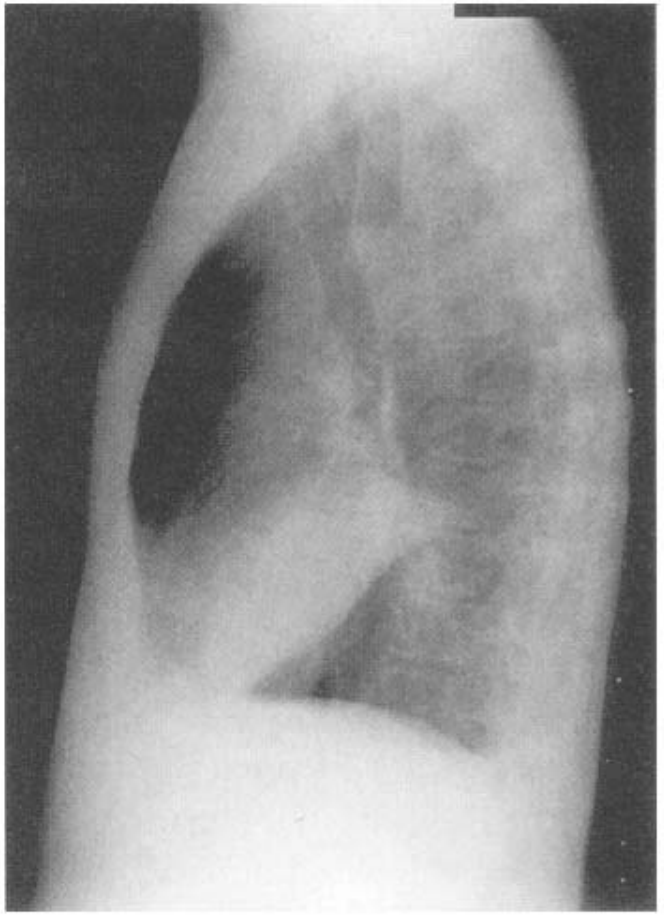
Pulmonary hemorrhage: bilateral





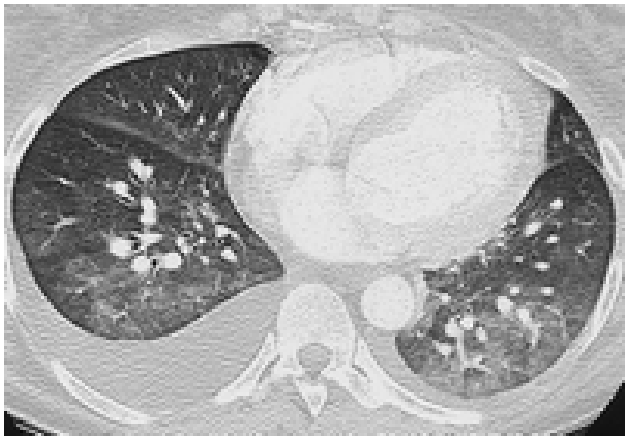
Unilateral pulmonary haemorrhage: R middle lobe consolidation

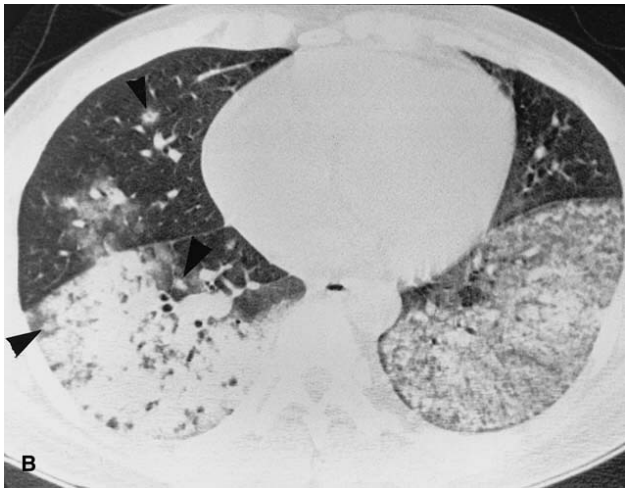


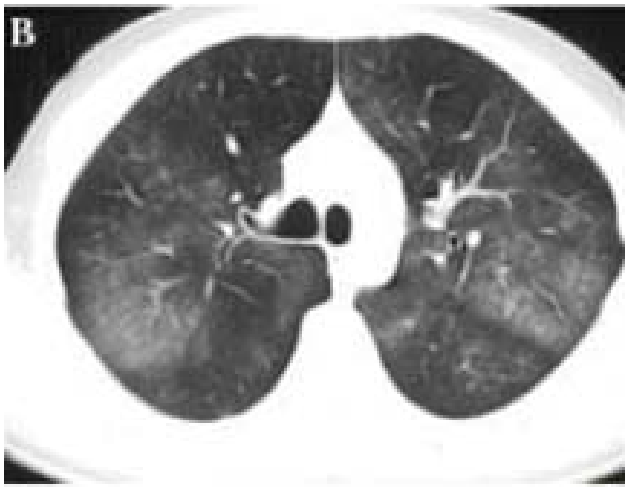


Pulmonary hemorrhage

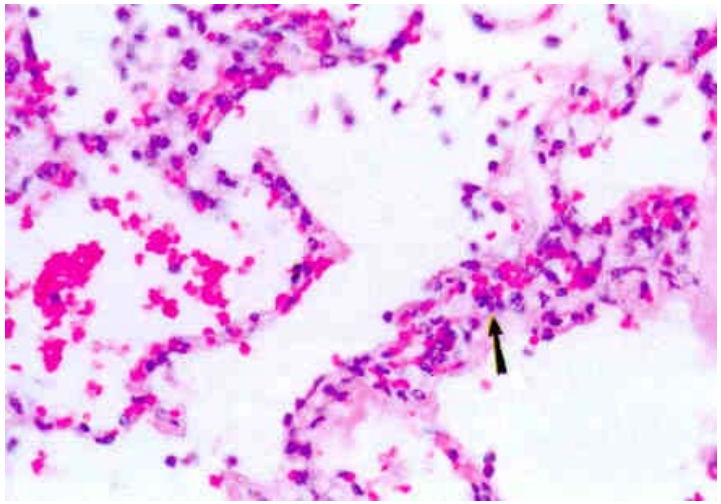
- CT findings:
  - Ground glass attenuation
  - Dense consolidation

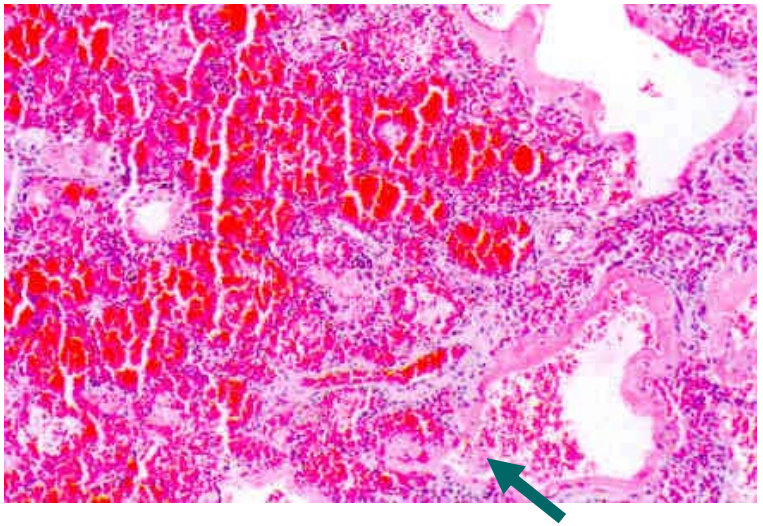






Pulmonary hemorrhage





- Interstitial infiltrate
- Scattered RCC in alveolar space

- Diffuse alveolar hemorrhage
- Capillaritis

Pulmonary hemorrhage

- Treatment
  - No prospective controlled trials
  - High dose corticosteroid alone may not be effective in severe cases
  - Pulmonary haemorrhage developed in 13 patients after large dose of corticosteroid had been given in treating lupus nephritis with nephrotic syndrome

- Liu MF et al. Clinical experience of 13 cases with severe pulmonary haemorrhage in systemic lupus erythematosus with active nephritis. Scand J Rheumatol 1998; 27

Pulmonary hemorrhage

- Plasmapheresis:
  - 3 active SLE patients with acute life-threatening pulmonary haemorrhage
  - 2 with recurrent bleeds despite treatment with cyclophosphamide and high dose steroid
  - Plasmapheresis → rapid clinical and radiographic improvement
  - 2 survived without long term lung problem
  - 1 died of sepsis after initial response to plasmapheresis

- Erickson RW et al. Treatment of haemorrhagic lupus pneumonitis with plasmapheresis. Semin Arthritis Rheum 1994

Pulmonary hemorrhage

- 7 patients with 11 episodes
- Prednisolone, 1-3mg/kg/d (n=2) or
  iv methylprednisolone 1g/d (n=9)

- ±oral cyclophsphamide 2-3mg/kg/d (n=7)
- Plasmaphoresis (n=5)
- 100% survival rates
  - May relate to unilateral involvement in some of the cases

Aberto S, et al. Chest 2000;118:1083-1090

Variables	Current Series	Abud-Mendoza et al[6]	Myers et al[7]	Schwab et al[8]	Zamora et al[9]	Barile et al[10]	Koh et al[11]	Liu et al[12]
Acute treatment, % of episodes								
CS	100	83	100	100	78	100	100	100
CYC	70	8	25	62	68	5	80	15
AZA	0	16	0	0	26	0	0	—
PAP	40	—	25	12	36	5	40	—
VENT	30	—	75	50	68	59	80	77
ABX	90	—	—	75	—	94	100	—
Survival, %	100	8	50	75	46	38	60	23
Mean AH-free Follow-up, mo	7.8	—	—	20	30	2	22	—
Range, mo	1–22	—	—	0.5–48	1–108	0.1–8	3–108	—

*CS = corticosteroids; PAP = plasmapheresis; VENT = mechanical ventilation; ABX = antibodies; see Table 1 for abbreviations.

Table 2—*Characteristics and Clinical Presentation of Patients with SLE-Associated AH (Current and Selected Case Series From 1985 to Present)***

Variables	Current Series	Abud-Mendoza et al[6]	Myers and Katzenstein[7]	Schwab et al[8]	Zamora et al[9]	Barile et al[10]	Koh et al[11]	Liu et al[12]
Patient characteristics								
Patients, No.	7	12	4	8	15	34	10	13
% of SLE cohort	1	1.6	—	1.4	3.7	5.4	1.4	4
Male/female, No.	1/6	0/12	3/1	2/6	5/10	2/32	2/8	1/12
Mean age, yr	31.1	23	29	37.8	30.1	34.5	26.5	26
Range, yr	19–44	16–40	13–51	17–54	19–44	—	13–44	10–50
Mean duration of SLE, yr	4.5	2	—	2.3	2.5	14.1	1.8	1.9
Range, yr	2 wk–19 yr	0.1–5	—	0.1–7	0–8	—	0–5	4 wk–5 yr
(+) anti-dsDNA, %	43	25	75	87.5	—	88	50	61
Presenting signs and symptoms of AH, % of episodes								
Hemoptysis	50	25	75	100	42	58	30	84
New radiograph infiltrates	100	83	100	87	100	100	100	100
Alveolo-interstitial	80	83	100	—	100	—	100	—
Lobar	20	—	0	12	0	—	0	—
Bilateral	80	—	100	—	100	—	—	100
Unilateral	20	—	0	12	0	—	—	—
Pleural effusions	30	—	—	—	37	—	—	—
Anemia	90	100	—	75	94	91	100	100
Dyspnea	100	25	87	73	—	100	—	100
Fever	80	25	25	100	26	—	90	54
Chest pain	30	—	—	—	—	—	20	—
Extrapulmonary signs and symptoms accompanying AH, % of episodes								
Renal-nephritis	70	41	—	62	93	32	40	100
Hematologic								
Leukopenia	30	50	—	—	0	—	20	23
Thrombocytopenia	20	—	—	—	31	—	—	61
AIHA	—	—	—	—	—	—	—	—
Skin-mucositis	0	41	—	62	73	47	70	38
Arthralgias-arthritis	10	16	—	62	15	44	30	15
Neuropsychiatric lupus	10	58	—	37	47	14	20	61
Low complements	83	25	75	100	—	—	70	84

*— = not stated; AIHA = autoimmune hemolytic anemia; see Table 1 for abbreviation.

Tacrolimus use in pulmonary hemorrahge

- 22-year-old woman
- SLE with fulminant pulmonary hemorrhage, mechanical ventilation
- Successful RX after continuous drip of **infusion of tacrolimus**, plasmapheresis and IVIG, corticosteroid

  - K Hoshi, et al. Clin Rheumatol 2004; 23:252-255

Case 2

- 34-year-old woman
- Admitted to hospital for digital necrotic hand lesions
- CT scans: right adrenal and liver focal necrosis
- Seizures (normal CT and MRI brain)
- Renal insufficiency
- Anemia and thromocytopenia
- Schistocytes and increased LDH levels
- Dx as **TTP**

- M Bortolati, et al. Autoimmu Rev 2009 8: 297-301

- Prednisolone + Daily plasma exchange using FFP as replacement fluid
- No improvement after 7 consecutive PE sessions

- Lupus anticoagulant +, high IgG aCL and anti-β2 glycoprotein I
- ADAMTS-13 values: normal
- Anti-ADAMTS-13 antibodies: absent

Catastrophic antiphospholipid syndrome (CAPS) with microangiopathic involvement

- Plasma exchange (PE) with using 4% human albumin as the replacement fluid
- iv heparin infusion
- Prednisolone 80mg/day

- Clinical condition and lab. parameters improved following 11 PE sessions
- Warfarin (target INR 2.0-3.0) + low dose aspirin
- Patient was discharged

Antiphospholipid syndrome (APS)

- Thromboembolic phenomena
- Obstetric morbidity
- Antiphospholipid antibodies
  - Anticardiolipin Ab
  - Lupus anticoagulant
  - Anti-β2 glycoprotein I Ab

Catastrophic APS (CAPS)

Classification criteria:

- multiorgan thrombosis developed simultaneously or in less than a week
- affecting at least 3 organs, systems and/or tissues
- histopathologic confirmation of small vessels occlusion in at least one organ or tissue
- Presence of anti-PL

- Asherson RA, et al. Lupus 2003;12:530-4

CAPS

- Represent less than 1 % of all patients with APS
- Approximately 60% of the catastrophic episodes are preceded by a precipitating event, mainly infections

CAPS

- May be completely missed
- High mortality rate (~50%), mostly due to cerebral and cardiac thrombotic involvement, infections and multi-organ failure

CPAS Registry

- Created in 2000 by the European Forum on aPL
- **250 patients**
- 70% are women
- Mean age of 37 yrs (range, 7-76)
- 46% primary APS
- 40% SLE
- 46% CAPS de novo (46.4%), without any previous hx of a thrombosis
- Previous manifestations: DVT, fetal loss, thrombocytopenia

S Bucciarelli, et al. Lupus 2009;18,905-912

CAPS

- Organs affected by the thrombotic event and the extent of the thrombosis
- Manifestations of the systemic inflammatory response syndrome (SIRS)

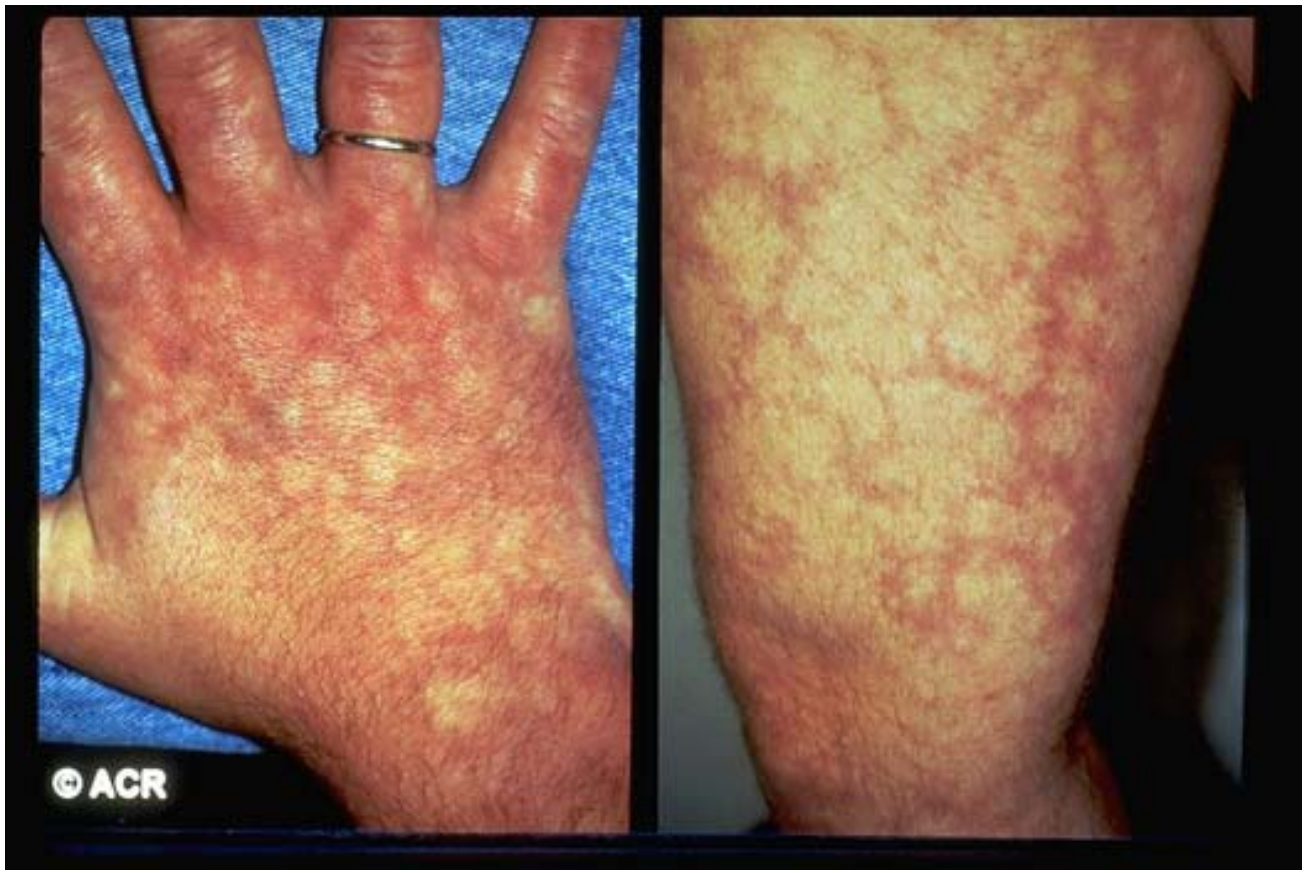
CAPS

- **Abdominal pain** due to intraabdominal thrombotic complications – most common presentation
  - Kidney, adrenal glands, splenic, intestinal and mesenteric or pancreatic vasculature
- Renal disease 70.6%
- Pulmonary complication: 69.3%
  - ARDS
  - Pulmonary embolism
  - Minority: pulmonary haemorrhage, pulmonary oedema and infiltrate

CAPS

- Cerebral manifestation 62%
  - Infarcts, encephalopathy, seizures or cerebral venous occlusions
- Skin complications 50.2%
  - Livedo reticularis, purpura and skin necrosis
- Cardiac problems 51.4%
  - Valve defect (mitral, aortic)
  - MI 25%

Livedo reticularis



CAPS

Other organs may be occasionally affected:

- Testicular or ovarian infarction
- Necrosis of prostate
- Acalculous cholecystitis
- Bone marrow infarction
- Oesophageal rupture
- Diant gastric ulceration
- Colonic ulceration
- Thormbotic pancreatitis
- Adrenal infarction

Systemic inflammatory response syndrome

Cytokines activation

- ARDS
- Cerebral edema
- Myocardial dysfunction

2003 International Consensus Statement on CAPS

Rx recommendation:

- Anticoagulants (AC)
- Corticosteroids (CS)
- Plasma exchange (PE)
- And /or IVIG

- **AC + CS** – most common therapy used
- **AC + CS + PE** and/or IVIG – 2nd most common regimen

- Therapeutic regimen combining anticoagulation, corticosteriods and plasma exchange has the best recovery rate (77.8%)

- Mortality rate
  - 53% (1992-2000)
  - 33% (2001-2005)

- iv heparin, 7-10 days
- Warfarin, INR ~3

- Corticosteroid:
  - for a minimal of 3 days, further Rx depending on patient's response

Role of Plasma exchange

- 4 patients with definite or possible criteria for CAPS
- All 4 had a favorable outcome
- All were treated with plasma exchange, which was promptly administrated **within the first 7 days of hospitalization**

- M Bortolati, et al. Autoimmun Rev 2009:8;297-301

Plasma Exchange

- Not only removes pathogenic antibodies but also clotting factors, cytokines and complement activation products contributing to the disease's pathogenesis

PE: the best replacement fluid ?

- FFP vs albumin solution
- FFP contains both:
  - Natural anticoagulants such as antithrombin III and protein C
  - Clotting factors
- Albumin
  - Could reduce transfusion/allergic reactions induced by plasma
- In 2 of the 4 cases, clinical picture worsen after FFP infusions; but they improved when PE using albumin solution as replacement fluid was administrated

- M Bortolati, et al. Autoimmun Rev 2009:8;297-301

Case 3

- M/61
- GP consultation for fever, dyspnea, myalgia in lower extremities
- CXR: interstitial changes in both lower lung fields
- Admitted to a hospital, given antibiotics
- Progressive dyspnea, rapid deterioration pulmonary interstitial change on CXR
- Methylprednisolone pulse therapy, transferred to other hospital

- Mod Rheumatol 2006; 16:55-57

- Aferbile
- Coarse crackles in both lung fields
- Normal dermatologic features
- Profound proximal muscle weakness in both lower extremites
- CPK 2540 IU/ml
- Anti-Jo 1 antibody 164 U/ml
- CXR: reticulonodular pattern in middle and lower lobes
- CT : ground-glass opacity
- TrBx: inflammatory changes in the interstitium infiltrated mainly with lymphocytes, without formation of hyaline membrane or granuolma, **consistent with NSIP**
- Muscle Bx: changes typical of myositis

Polymyositis with interstitial pneumonia

- Prednisolone 60mg and Cyclosporin A 100mg
- Improvement in CXR and CPK values
- 14 days after RX:
  - Subcutaneous emphysema
  - CXR and CT: pneumomediastinum
- Gradual disappearance
- Interstitial pneumonitis and muscle symptoms improved on treatment
- Discharged with prednisolone 20mg and CSA 100mg on the 30th day following admission

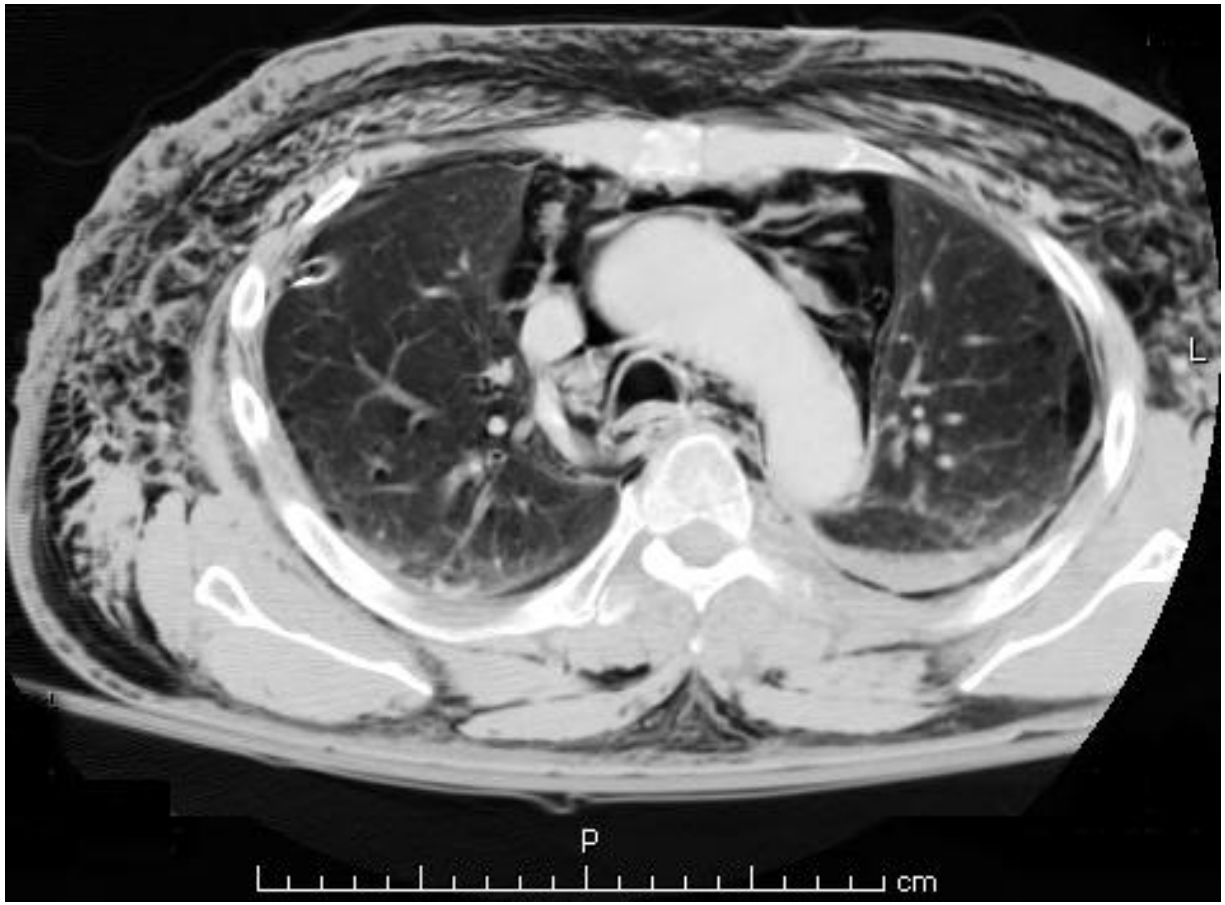
Polymyositis/dermatomyositis

Respiratory manifestations:

- As high as 45%
- Infection
- Aspiration pneumonia
- Malignancy
- Drug-induced pneumonitis
- Respiratory muscle weakness
- **Acute or chronic interstitial lung disease**

Interstitial pneumonitis in DM and PM

- Frequency 5-42%
- Rapid progression had often been reported in DM but only occasionally reported in PM
- Pneumomediastinum is rare complication that had been reported mainly in DM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Pneumomediastinum in interstitial pneumonitis

- A rare complication
- Had been reported mainly in DM
- Tends to be resistant to conventional therapy
- 56% of those cases were fatal [1]
- The result of air bursting from the alveolus and spreading through the neighboring tissues along the pulmonary vessels toward the mediastinum

[1]Satomi k, et al. Nihnon kokyuki Gakkai Zasshi 1998;36:984-8

PM/DM-associated lung disease: analysis of a series of 81 patients

- Retrospective study
- 50 (61%): some type of pulmonary involvement
- **32 (39%) had ILD**
- 15 (47%) were ASA +ve:
  - anti-histidyl-tRNA synthetase (anti-Jo-1)
- **5** with devastating acute interstitial pneumonia with **pneumomediastinum** and an unfavorable prognosis
  - 3: had died by 3 months after ds onset
  - 2: lung transplantation ; died 1 & 6 months after transplanation

A Selva-O'Callaghan, et al. Lupus 2005:14;534-542

- Abnormal bronchoalveolar lavage fluid in 19 patients

  (normal reference <15% lymphocytes, <5% neutrophils)

  - Neutrophil-predominant in 5
  - Lymphocyte-predominant in 14

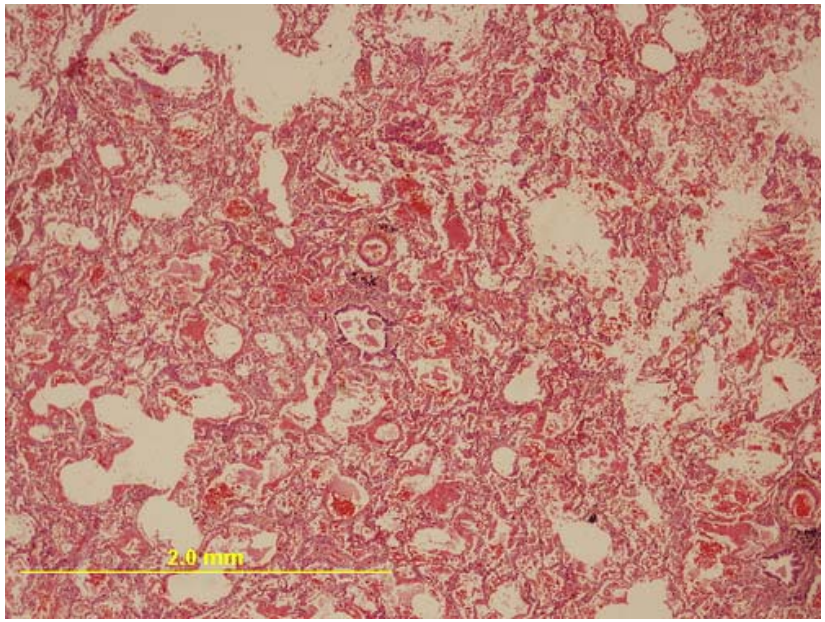
- Main pathological pictures:
  - Nonspecific, irregular fibrotic pattern associated with homogeneous interstitial alveolar thickening due to mild chronic inflammation, edema, interstitial fibroblast proliferation and pneumocytic hyperplasia
  - 5 death cases: **diffuse alveolar damage**

Survival in patients with ILD

- **Acute ILD with pneumomediastinum**: poorest survival time
- Anti-histidyl-tRNA synthetase (anti-Jo-1) +ve
  - Significantly associations with for ILD
  - Not related to mortality
- In ILD group, a trends to longer survival in ASA+ patients
  - 5-yr mortality was higher in ASA (-) than ASA (+) patients (5 out of 17 versus 0 out of 15)

Pneumomediastinum in 5 patients

- Subcutaneous emphysema
- Histological study: diffuse alveolar damage
- None with Anti-Jo 1 or ASA



Rx of ILD in DM and PM

- Initial Rx: Prednisolone 1mg/kg/day
- Interstital lung disease deterioration (chronic course with progressive deterioration of lung fx)
  - Azathioprine in 14 (1-2mg/kg/day)
  - Cyclosporine in 3 (3-5mg/kg/day)
  - Partial remission in **17 patients**

- Rapidly progressive course in **5 patients** and died
  - Prednisolone (1mg/kg/day) and pulse (1g/d for 3 day)
  - Cyclophosphamide (0.7g/m^2)
  - Cyclosporin A (5mg/kg/day)
  - Tacrolimus (3mg/12 hours)
  - IVIG (2g/day for 2 days)
  - Plasmapheresis in 2 cases

- **6 patients**
- cyclophosphamide pulses (0.5-0.7 g/m^2) for 6 months + prednisolone (1mg/kg/day)
- All with improvement in FVC at 6 months vs baseline

- Monthly pulses of cyclophosphamide
- 10 out of 36 PM/DM patients with ILD resulted in improvement or resolution of lung involvement in half the patients

Marie I, et al. Arthritis and Rheum 2002; 47: 614-622

Case 4

- F/51
- Menopausal, taking HRT
- Lower limb edema x 6 months; progressed to hands and face, generalized pain
- Features of systemic sclerosis localized to face, hands; pain in multiple muscles and joints, Raynaud's phenomenum

- HRT was discontinued and prednisolone was commenced at 1 mg/kg/d

- 4 weeks later, hospitalization for dyspnea, hypertension (240/120), headache, vomiting
- Hb 10.2, platelet 80
- Urea 29.8, Cr 329
- Urine: RBC and RBC casts
- ANF 1:640, dsDNA -ve, anti-ENA –ve

- Dx: **scleroderma renal crisis**

- Catptopril 6.25-12.5mg every 8 h, increased to 50mg tds

- Renal bx:
  - Initimal thickening and fibrosis of blood vessels and glomeruli
  - No evdience of inflammation or GN

Renal disorders in Systemic Sclerosis

- Scleroderma renal crisis (SRC)
- Chronic kidney disease
- Inflammatory renal disorder
  - Glomerulonephritis
  - Vasculitis
  - Interstitial nephritis

SRC

European cohort study[1]:

1996 adult SSc patients, SRC present in:

- 12% of diffuse systemic sclerosis patients
- 2% of limited systemic sclerosis patients

[1] PennH, et al. QJM 2007;100:485-494

Clinical presentation

- Accelerated phase hypertension
- Hypertensive retinopathy
- Hypertensive encephalopathy
- Non-nephrotic range proteinuria and hematuria
- Progressive renal impairment
  - Renal failure often occurs over weeks rather than days
- Microangiopathic hemolytic anemia
  - Significant coagulopathy is rare

Histology

- Characteristic lesion:
  - initimal and medial proliferation with luminal narrowing typically occurs in arcuate arteries
- Indistinguishable from changes of accelerated HT
- Fibrinoid necrosis and thrombosis are also common

Pathological findings

- **Acute Vascular changes** (mucoid initimal thickening and thrombosis) are associated with poorer outcome
- Half of those with acute changes require permanent dialysis, compared those with 13% without
- The index of chronic damage did not correlate with renal prognosis in SRC, reflecting the importance of vascular insult

Risk factors for SRC

- 65-86% of SRC occurs in patients with **diffuse SSc**
- **Early phase** of dcSSc
- **Rapidly progressive skin ds** (found in early phase of dcSSc)
- Up to a quarter of patients with SRC, the Dx of SSC is made at the time of the renal presentation
- Estimated median duration of SSc at SRC Dx is mere 8 months
- 66% of patients with SSc develop SRC within a year of Dx, rising to 86% at **4 years**
- Limited SSc typically develop SRC later in SSc disease course

Risk factors for SRC

- 60% of patients have received **corticosteriods prior to presentation**
- A recent hx of **high dose corticosteriod** use (e.g. prednisolone or equivalent at > 15mg/day) preceded SRC Dx

- Although **chronic steroid use** may not be associated with additional risk, a greater number of patients who recently received low-dose corticosteroids developed SRC compared with controls
  - Odd ratios 17 (1 month exposure) and 24 (3 month exposure)

- Patients with SSc most likely to receive corticosteriods are those with early dcSSc, who are the same patients at greatest risk of SRC

Risk factors for SRC

- **ANA speckled pattern** positive status
  - Occurs in 60% of patients with SRC in one cohort
- **SSc-specific anti-RNA polymerase antibodies (I and III)**
  - In 59% of patient with SRC in one cohort

- Anticentromere Ab is relatively protected from developing scleroderma renal crisis

- **ACE inhibitors in SRC**
- 1 yr survival, 15% without and 76% with ACEIs
- Target:
  - ↓ systolic BP by 20mmHg and diastolic by 10mmHg per 24 hour
  - To normal range
  - Avoiding prolonged periods of hypotension
- Captopril, short half-life, easiest to titrate BP initially but longer acting ACEI are usually more convenient in the long term

- **Angiotensin II receptor blocking agents** can be safely added if once ACEIs are ineffective; but may not be sufficient to control disease in the absence of ACEI therapy

- Alpha-blocker, Ca antagonists

- iv prostacyclin (0.2-2ng/kg/min) [1]
- Increase renal perfusion
- Help to normalize BP
- Continuous infusion during hypertensive phase of the renal crisis, with discontinuation when renal function improves or at initiation of dialysis

[1]Scorza R, et al. J Rheumatol 1997;24:1994-1948

- **Plasma exchange**
  - in profound microangiopathic haemolytic anemia
- **Renal support with hemodialysis**
  - ~ two-thirds of patients with SRC presenting to a specialist centre will require renal replacement therapy
  - ~ half of them will recover sufficiently to discontinue dialysis
  - Poor outcome:
    - low BP at presentation,
    - presence of acute vascular changes on renal bx
    - older age if dialysis is required

- Renal recovery can occur more than 24 months after the crisis
- Renal transplantation
  - consider if dialysis continued for 2 yrs

End

Thank you