Idiopathic Pulmonary Fibrosis

(IPF)

How we could do better

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Medical Update Group at UoM

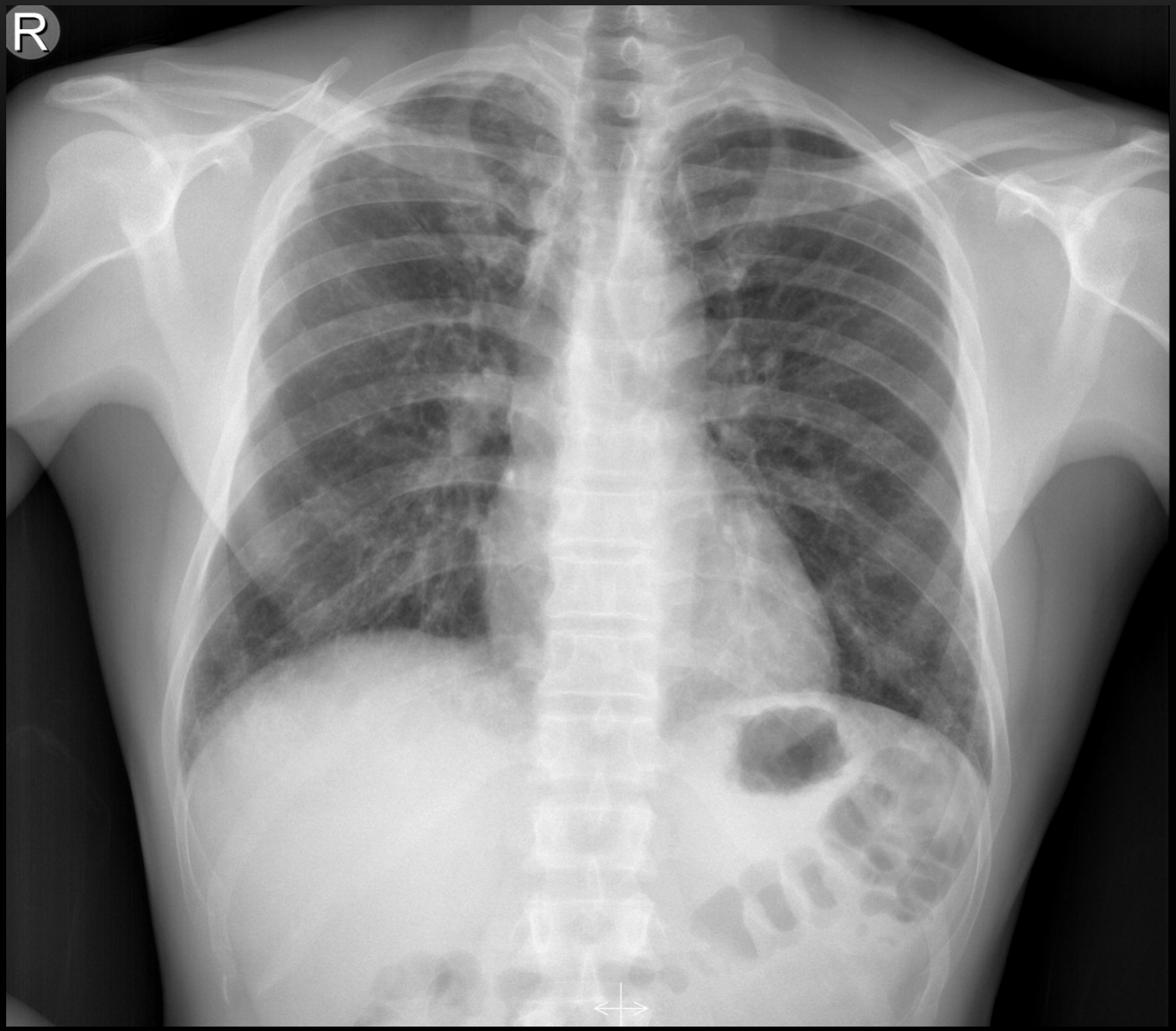
PATIENT CASE – MR. A. H, 65 YR (CONTRACTOR), EX-SMOKER, 15/DAY

- 2 year h/o cough – treated as asthma
- SOB 500 metres
- Chest X-ray only 2 weeks ago
- No known asbestos/ birds/ drugs
- No arthropathy/ skin/ eyes

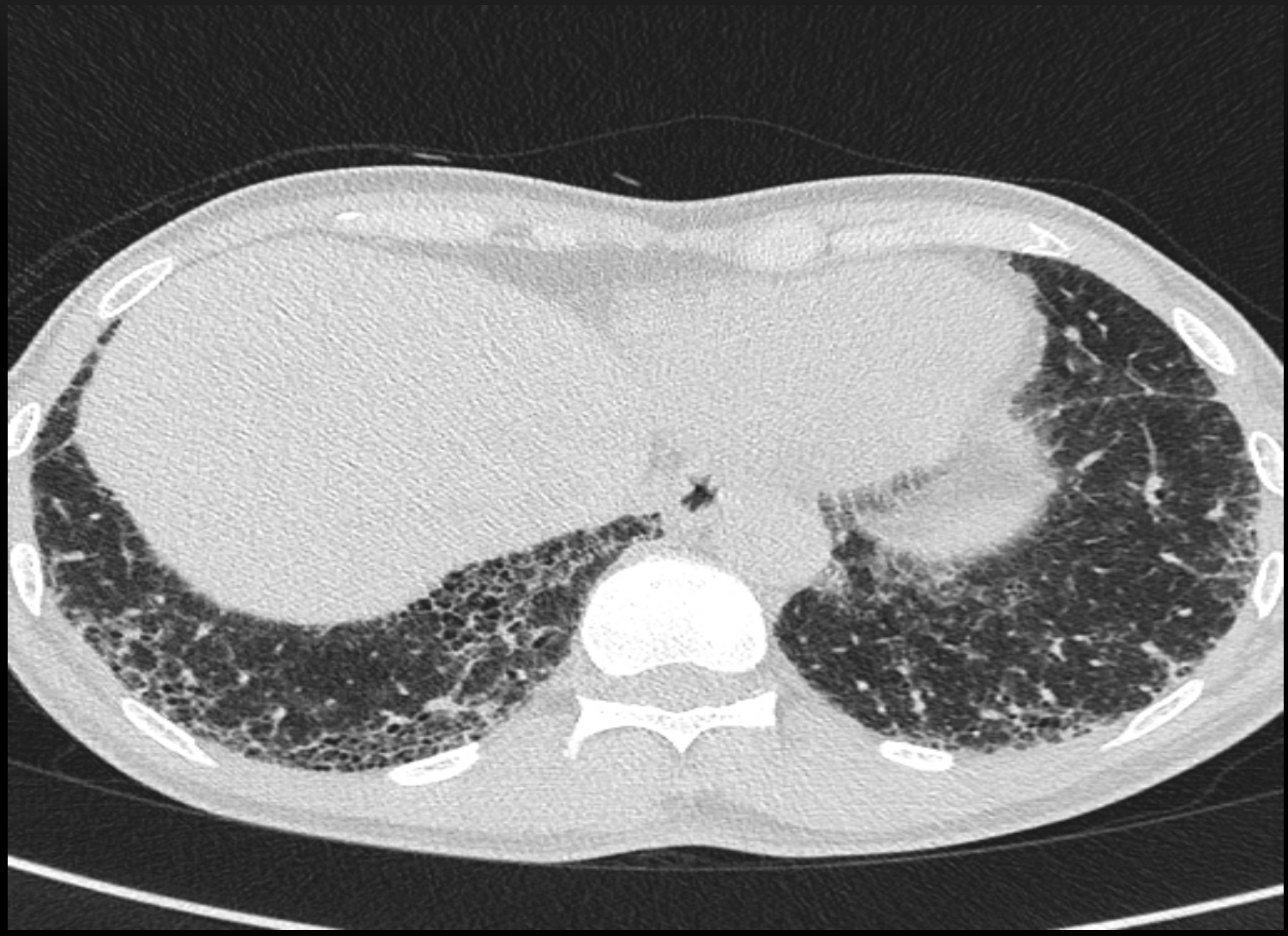
IPF CASE

- O/E
- Clubbed
- Showers of inspiratory crackles 2/3 up both lungs
- VC 55% DLCO 33%
- Show CXR and HRCT

FIRST CXR



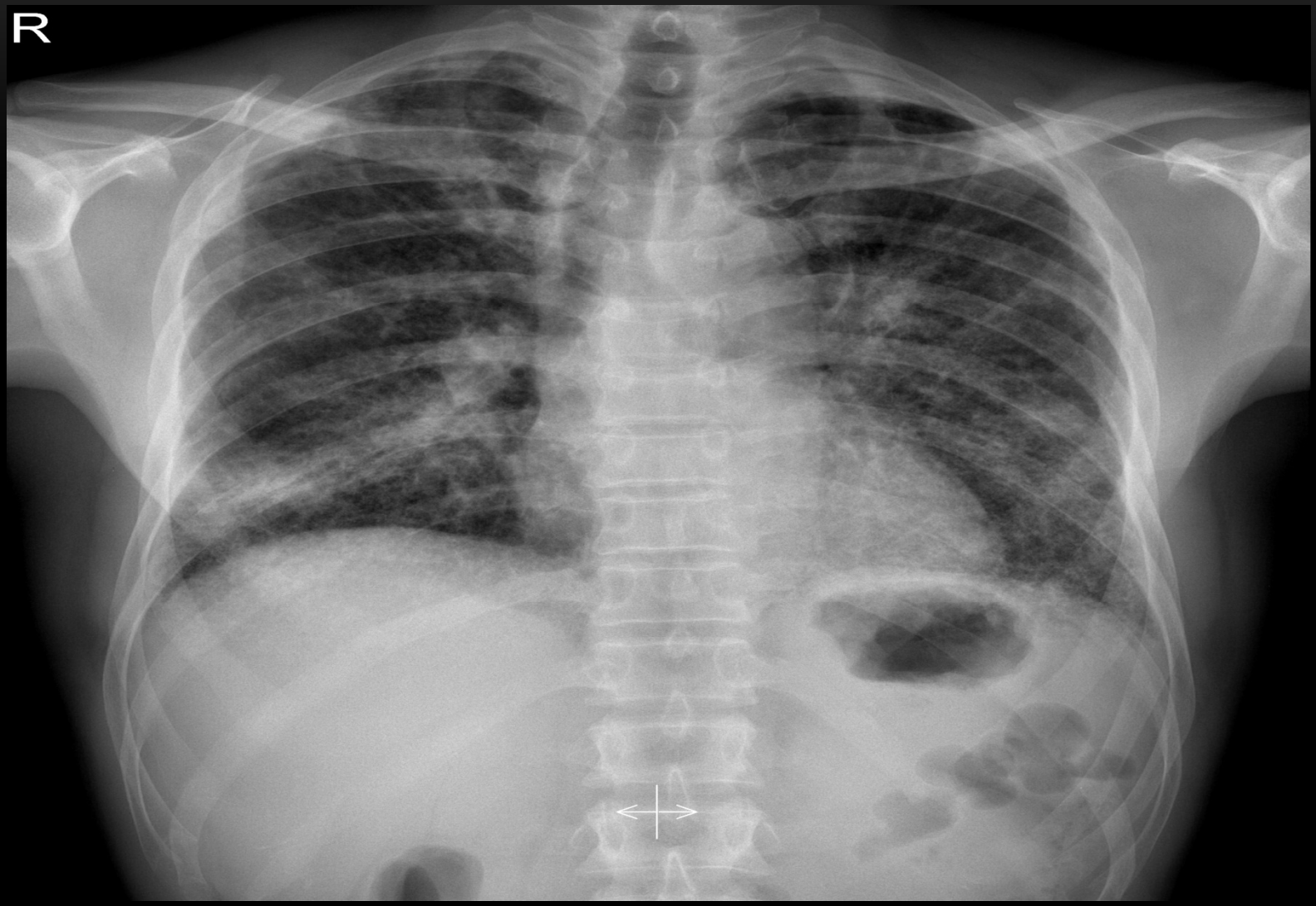
FIRST HRCT



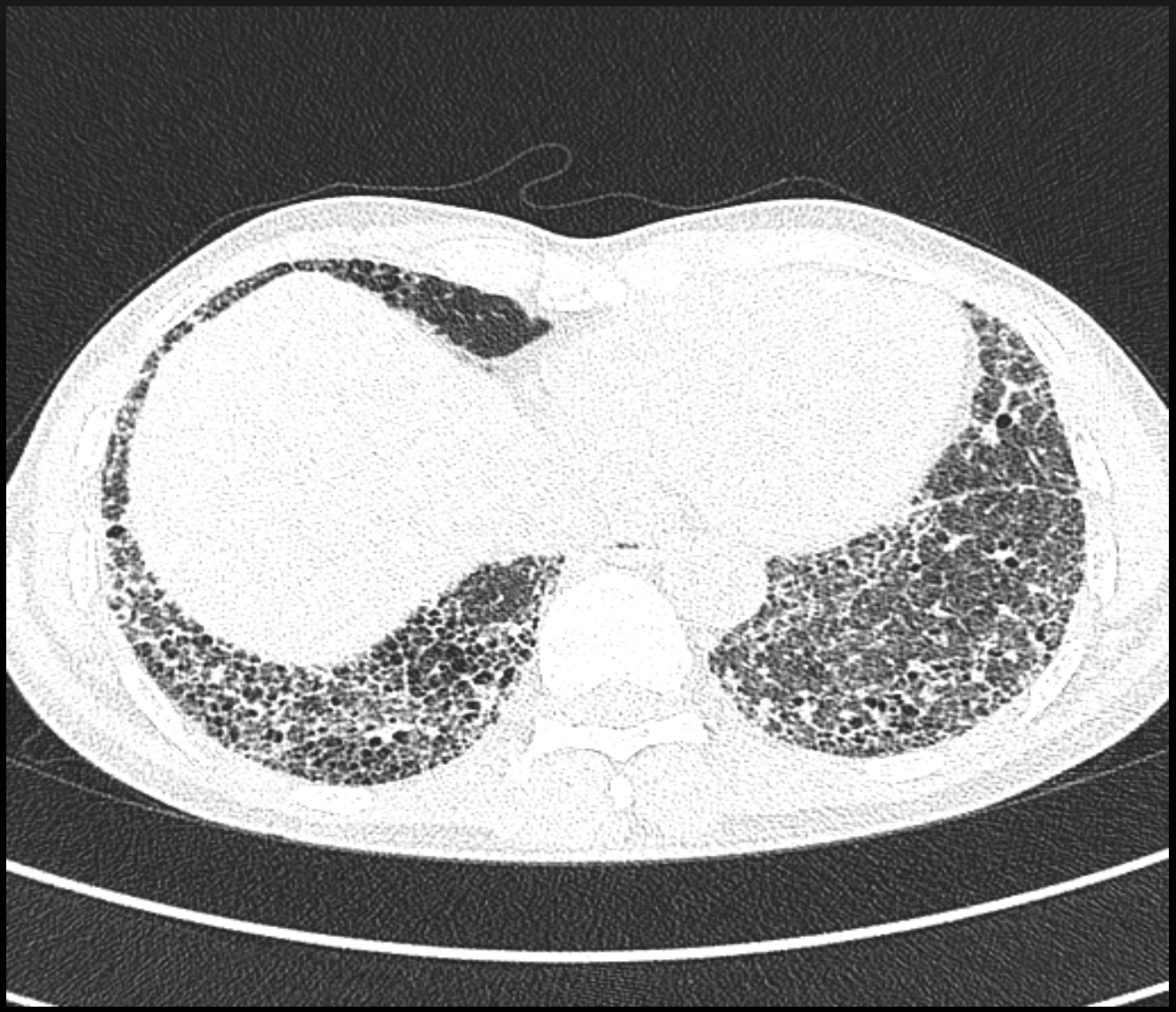
TREATMENT

- Oral corticosteroids
- Pirfenidone was not available then
- Slow worsening
- Admitted with exacerbation and died 3 years after presentation

TERMINAL CXR



TERMINAL HRCT



DIFFUSE LUNG DISEASE

Table 5.1 *Classification of diffuse lung disease*[a]

Primary disease-related DLD
Amyloidosis
Chronic aspiration
Chronic infection
Eosinophilic pneumonia
Inflammatory bowel disease
Langerhans' cell granulomatosis
Lipoid pneumonia
Lymphangioleiomyomatosis
Malignancy (lymphoma, metastatic)
Primary biliary cirrhosis
Pulmonary alveolar proteinosis
Sarcoidosis
Viral hepatitis

Environmental exposure-related DLD
Pneumoconiosis (inorganic substances)
Asbestos
Beryllium
Hard metals
Polyvinyl chloride
Silica

Hypersensitivity pneumonitis (organic substances)
Bagassosis
Bird fancier's lung
Ceramic tile lung
Chicken handler's lung
Detergent worker's lung
Farmer's lung
Fishmeal worker's lung
Goose down lung
Humidifier lung
Miller's lung
Mushroom worker's lung
Woodworker's lung

Drug-induced DLD
Amiodarone
Angiotensin-converting enzyme inhibitors
Antibiotics (cephalosporins, nitrofurantoin, ethambutol)
Chemotherapeutic agents (bleomycin, alkalating agents, methotrexate)
Cocaine
Dilantin
Radiation
NSAIDs

Collagen vascular disease-associated DLD
Ankylosing spondylitis
Polymyositis/dermatomyositis
Rheumatoid arthritis
Scleroderma
Sjögren's syndrome
Systemic lupus erythematosus

Idiopathic interstitial pneumonia
Acute interstitial pneumonia
Cryptogenic organizing pneumonia
Desquamative interstitial pneumonia
Idiopathic pulmonary fibrosis
Lymphocytic interstitial pneumonia
Non-specific interstitial pneumonia
Respiratory bronchiolitis-associated interstitial lung disease

[a]This is only a partial list of causes of DLD.

DIFFUSE LUNG DISEASE
The "Big 5"

PRIMARY
Disease-related, i.e.
SARCOID,
MALIGNANCY

ENVIRONMENTAL EXPOSURE
i) Asbestos
ii) Hypertensitivity pneumonitis (Wood workers)

DRUGS
Amiodarone
IVDA

IDIOPATHIC INTERSTITIAL PNEUMONIAS
- IPF DIP
- NSIP LIP
- COP RB-ILD
(Smokers)

COLLAGEN
Vascular disease SLE, RA, Sjogren's

EARLY DIAGNOSIS OF IPF

- Severe morbidity
- High mortality at 5 years
- Effective medical treatment now available but proven value when VC > 50%

IPF

- INSIDIOUS ONSET
- Dry Cough
- Slowly worsening dyspnoea

SIGNS

1. “VELCRO” inspiratory basal crackles
2. Digital clubbing
3. No signs of heart failure

INVESTIGATIONS

1. RADIOLOGY

- CXR & HRCT

2. Blood tests (As per clinical status)

- ESR, CRP, FBC, Diff, Urea/Creatinine
- Urine exam
- Autoimmune/ Vasculitis serology (Not routine)

3. PULMONARY FUNCTION TEST (VC)

4. ECHOCARDIOGRAM, ECG

LUNG BIOPSY OR NOT?

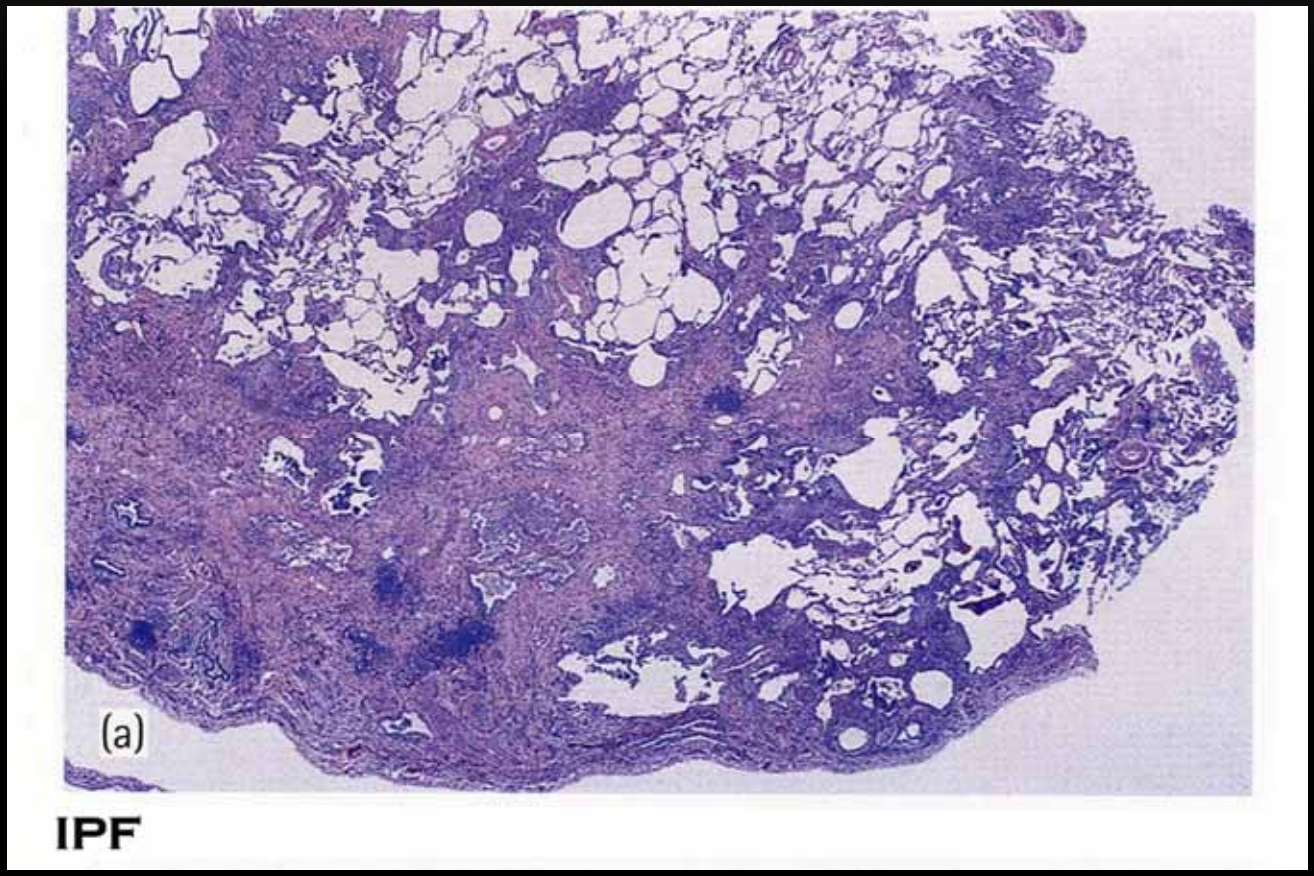
- HRCT appearances will be crucial (IPF or not IPF)
- If “classical” IPF – No need for steroids
- If not IPF and no other clues, then open lung biopsy if patient fit enough
- Show histopathology examples

WHY BIOPSY?

- STEROID responsive or not
- Avoid needless steroid
- Use IPF – Specific drugs, i.e:

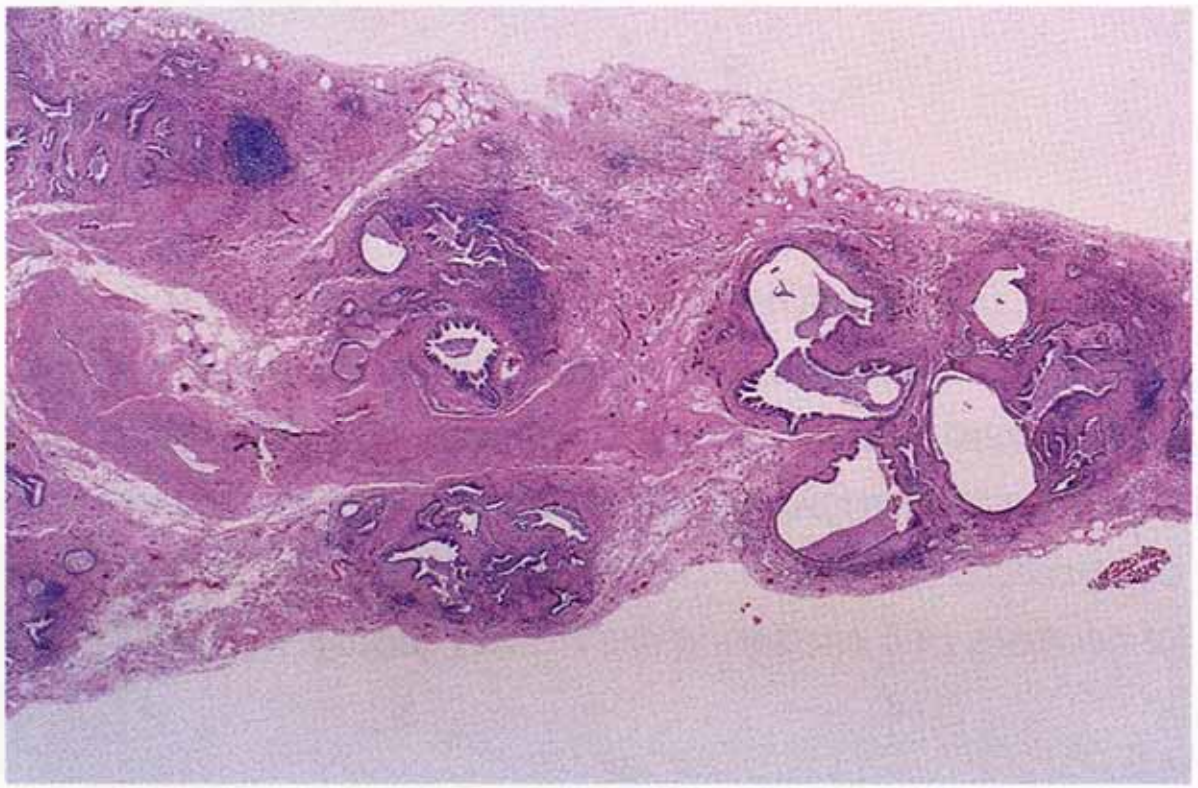
- Pirfenidone
- Nintedanib

IPF - HISTOPATHOLOGY



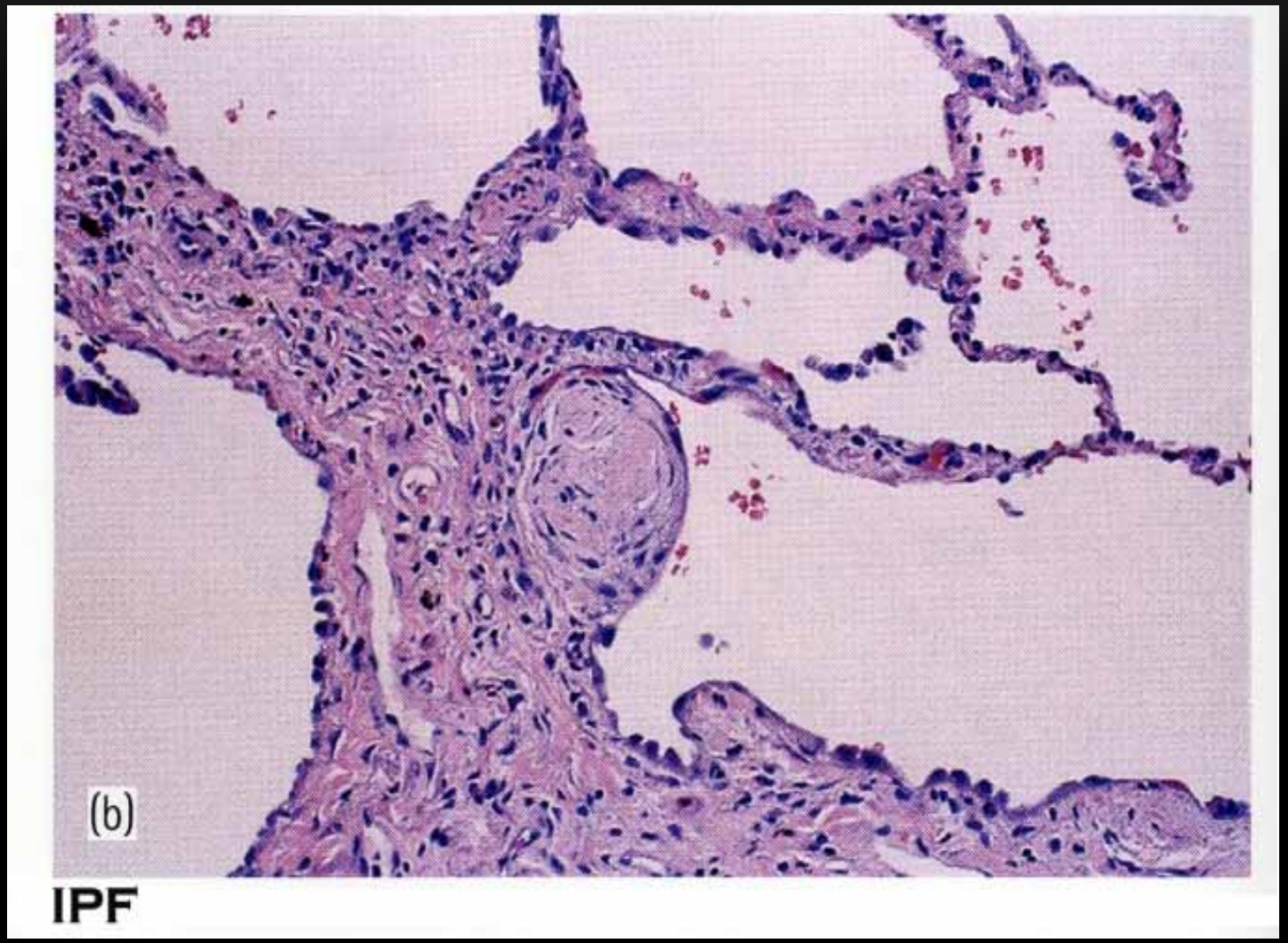
IPF

IPF – HONEYCOMB LUNG

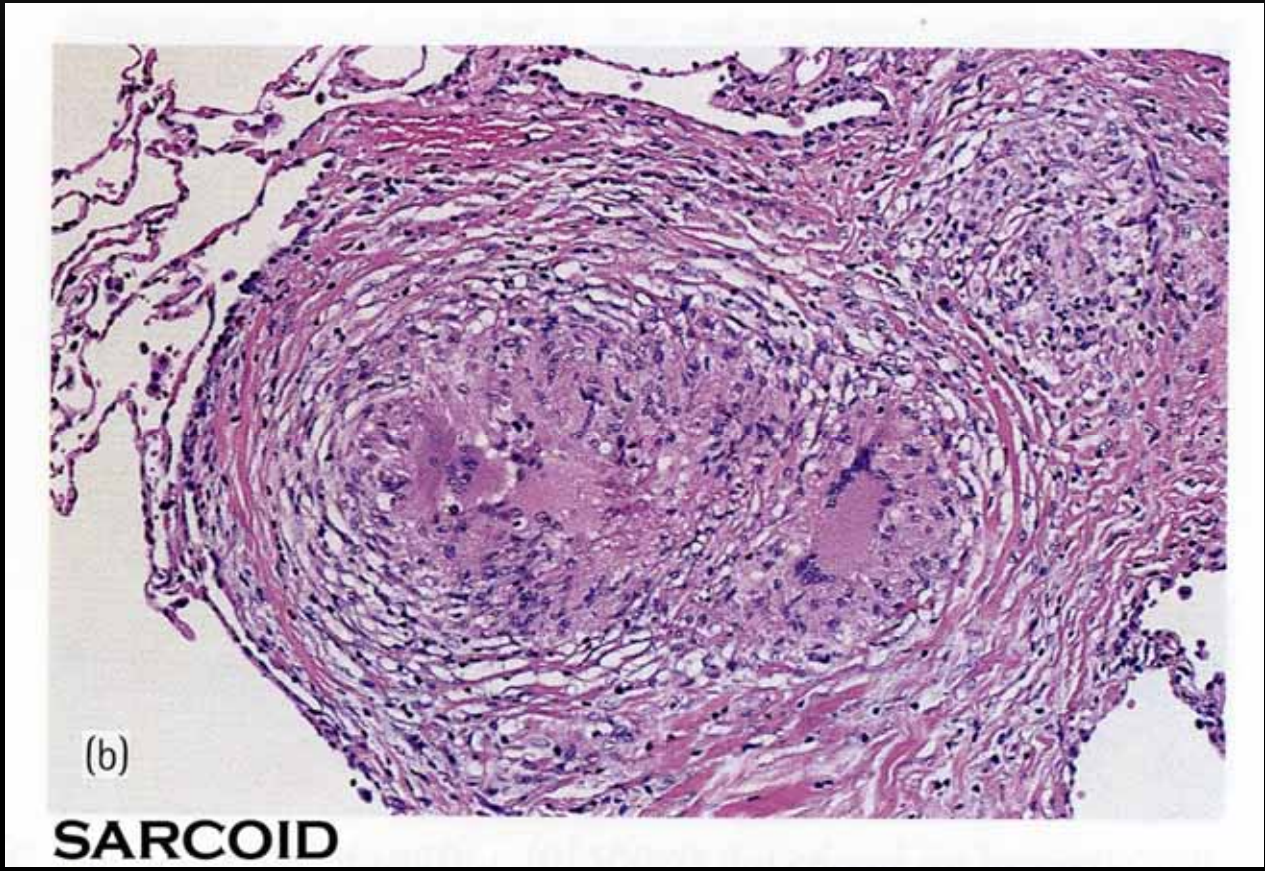


IPF

IPF – FIBROBLASTIC FOCUS



SARCOIDOSIS



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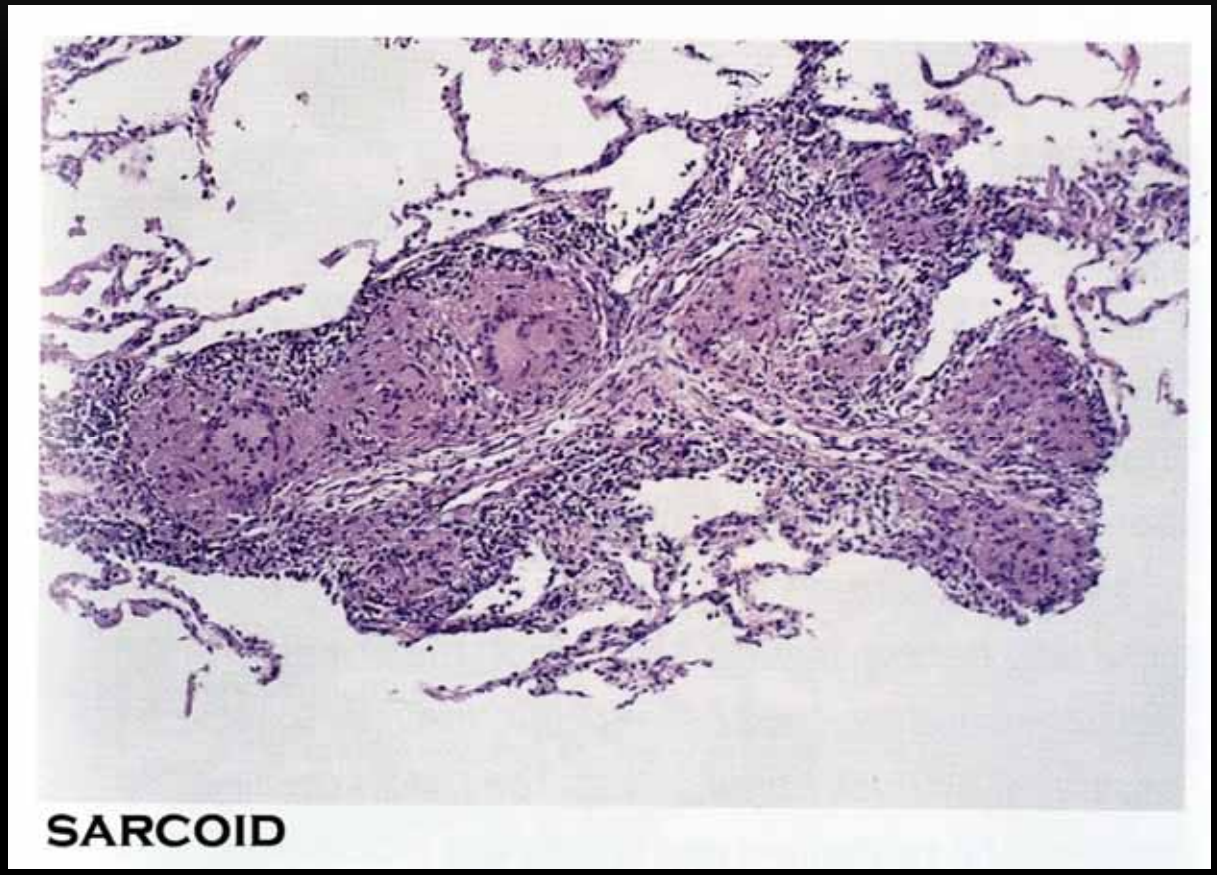
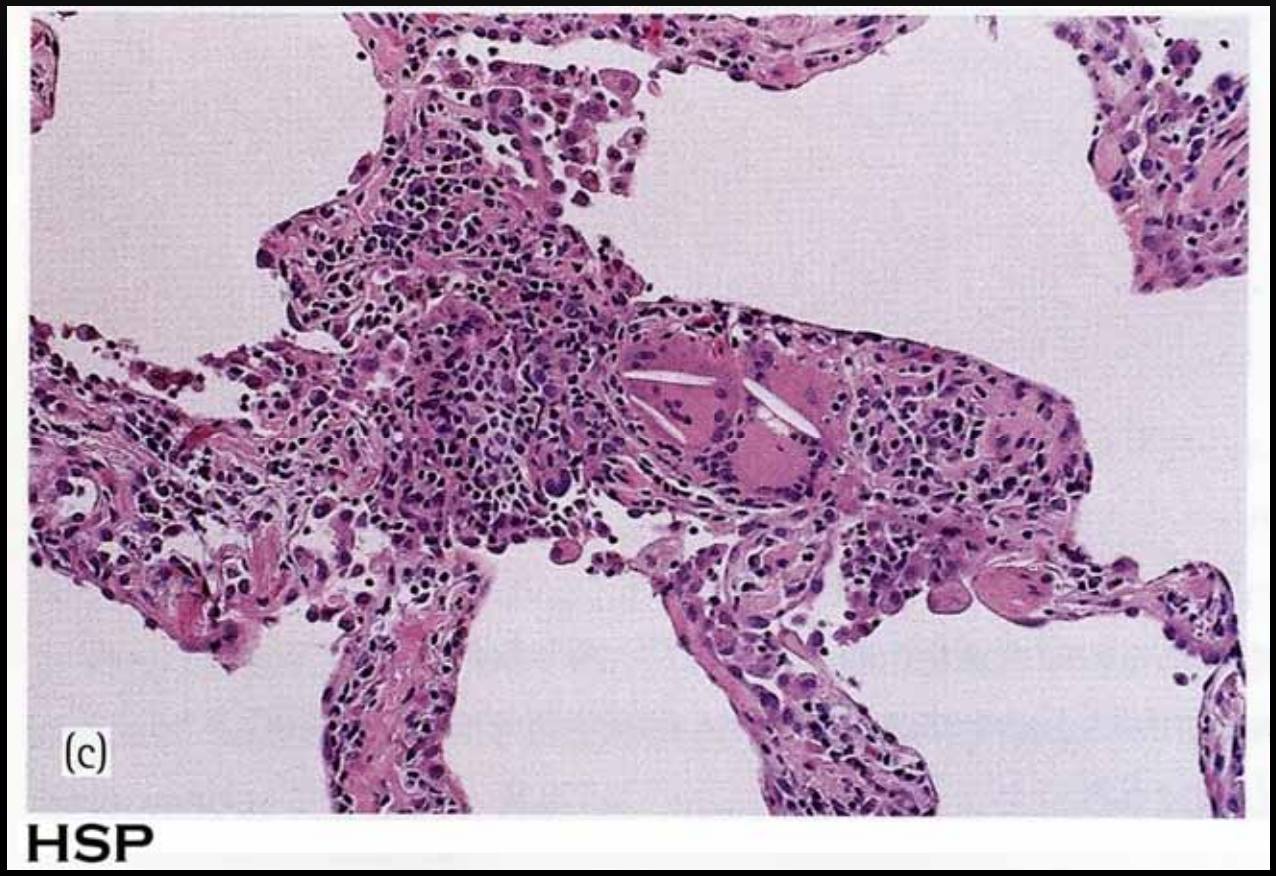


HYPERSENSITIVITY PNEUMONITIS



TREATMENT OF IPF

1. Specific

- Pirfenidone
- Nintedanib

2. Non specific

Home Oxygen

PPIs

Anti tussives

Vaccines

? Inhaled steroids

? Long term antibiotic

PULMONARY TRANSPLANTATION

- The definitive treatment (Not here yet)
- Beware human organ trafficking

TAKE HOME MESSAGE(S)

1. Spot early – Basal Crackles

- Think of IPF
- Basal crackles not always = Heart Failure

2. Early HRCT

+ Do few basal lung cuts in those > 55years undergoing Abdominal CT

3. Early referral to Pulmonary Specialist