FAKULTNÍ NEMOCNICE OSTRAVA

# Acute respiratory failure : a case report

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# Initial ED presentation

- 45 yr old, caucasian man, former hockey player
- Moderate dyspnoea, right hemithorax inspiratory pain
- 3 weeks prior to ER: joint swellling, fever, muscle weakness, unspecified fatigue
- ½ yr prior to ER: post-exertional dyspnoea
- Unremarkable family history, medication 0, no enviromental/occupational risk
- Physical examination: velcro-like crackles, TT=37.2 °C, $pO_2$=90%, mild proximal muscle weakness
- Lab.: ↑liver enzymes (AST 17x), creatinkinase (20x) , myoglobin (12x), ↑WBC ($19*10^9$,neutrophilia),CRP (2x)

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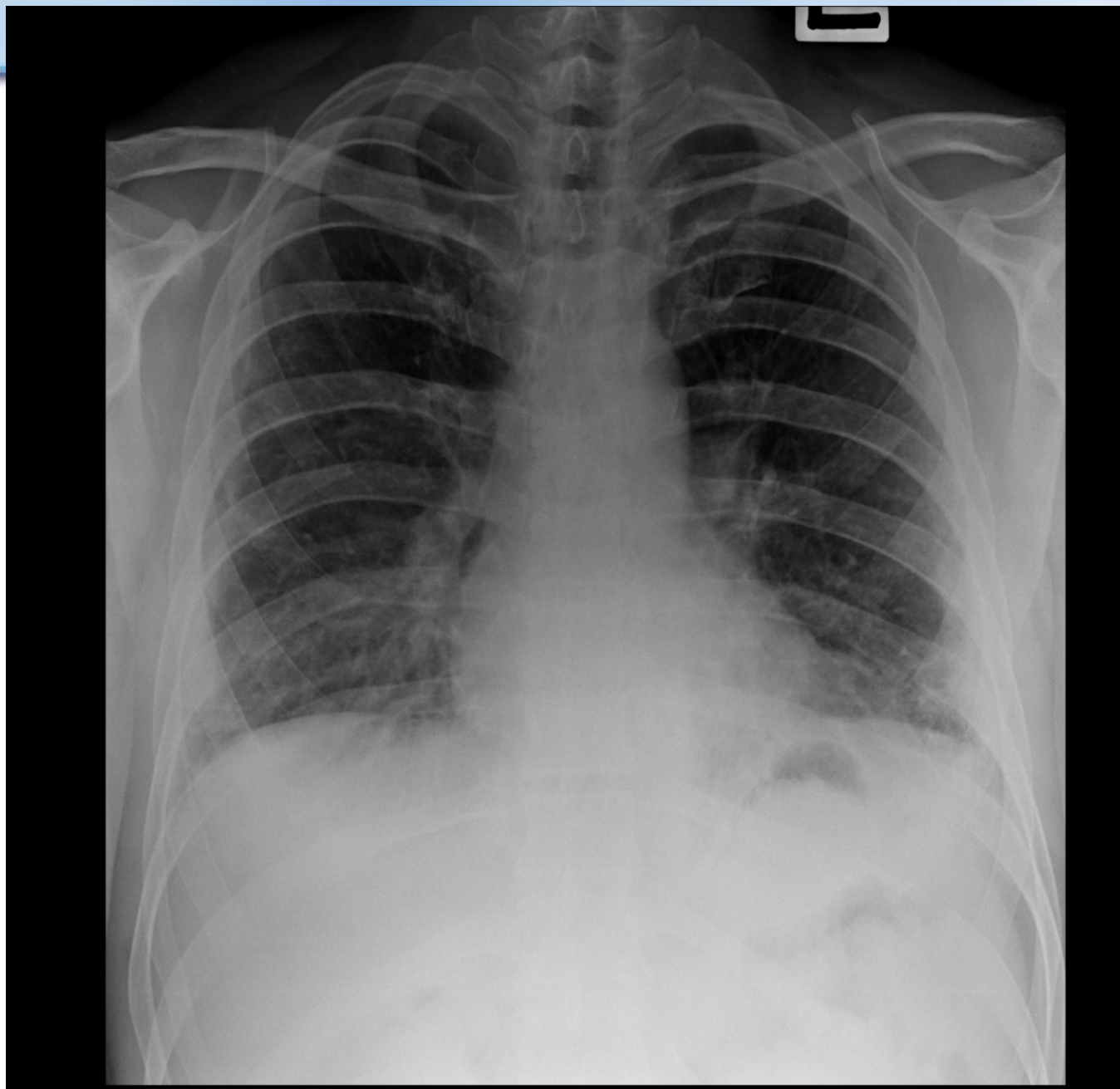
# Chest x-ray

Right basal pneumonia?

ATB



# Connective tissue dissease?

- ELISA IgM RF (127.37 IU/ml, N 1-25), mild + anti-Ro-52 (Western blot)
- Neurological exam: decreased muscle strenght (proximal 3/5, distal 4/5)
- Electromyography:signs of a diffuse myopathic pattern
- no inflammatory infiltration from muscle biopsy
- myositis? Solu-medrol pulse therapy (cumulative dose 3g)
- After 2 weeks: decreased levels of WBC, CK, CRP, myoglobin + subsiding fatigue/muscle weakness

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# Dyspnoea etiology?

- Saline induced sputum:negative both for bacteria and fungi.
- A full panel of respiratory viruses serology analysis proved noncontributory
- ECHO: no signs of systolic/diastolic dysfunction or valvular disease
- Oncomarkers: AFP 13 U/ml (range:0-11), CA 19-9: 3.8 U/ml (range:0-37), CEA 11.7 ug/l (0-5), **NSE 30.6 ug/l** (range:0-12.5), PSA 0.24 (range: 0-5), SCCA 0.4 ug/l (range 0-1.5)
- Spirometry: VC max=**1.2 l**, FEV1=**1.1 l**, Tiffenau index= 0.95

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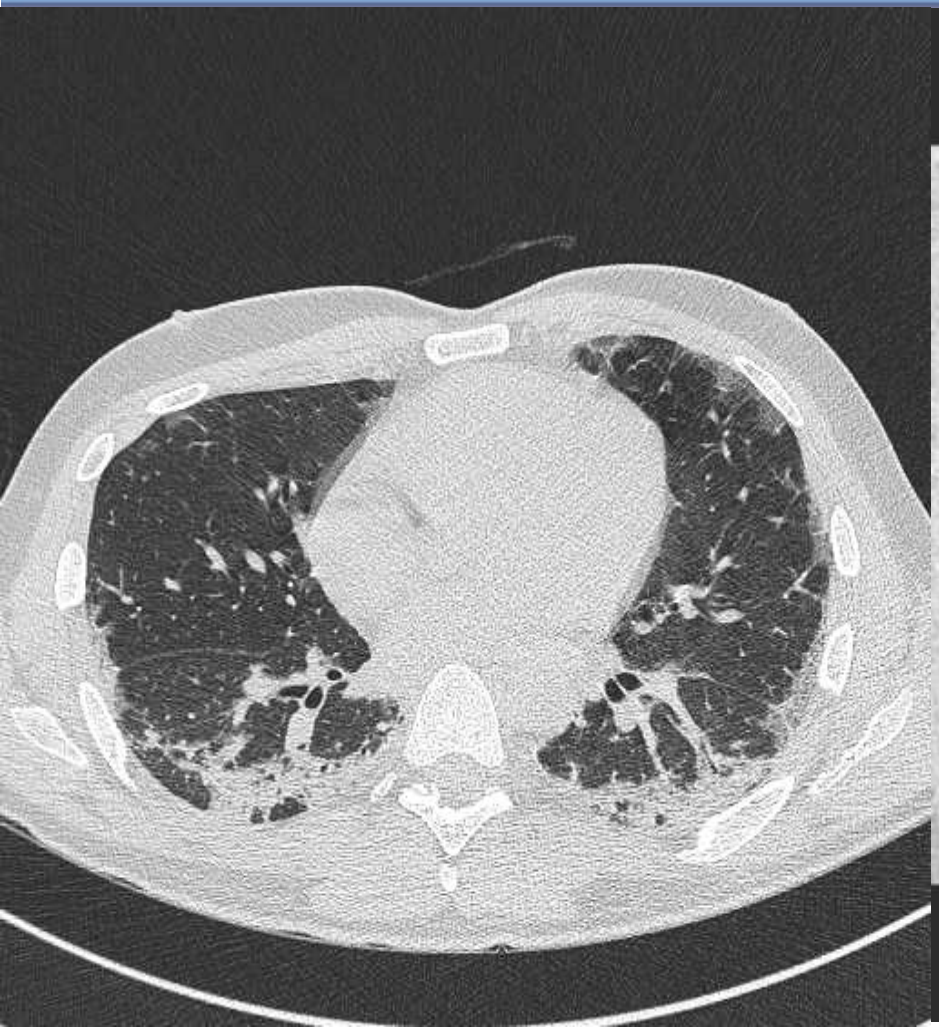
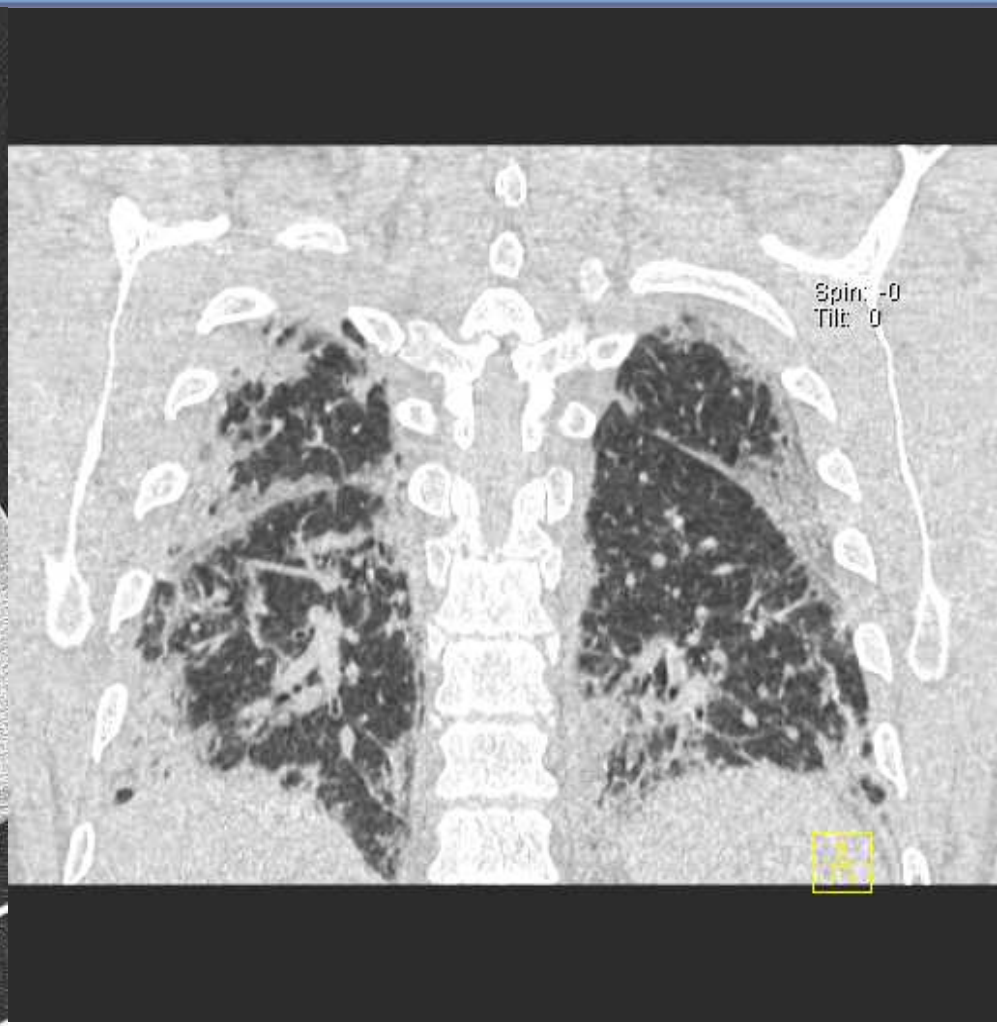
# High resolution computed tomography



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# New diagnosis: Idiopathic pulmonary fibrosis

- 2 major (abnormal pulmonary function+bibasilar reticular abnormality) ATS/ERS criteria were satisfied
- 2 minor (insidious onset of exertional dyspnoea + duration > 3 months) ATS/ERS criteria were satisfied
- BAL (after corticotherapy): Macrophages: 21,7%, **Granulocytes: 65%,** Lymphocytes: 11,9%, Eosinophiles 0,7%.
- Imunophenotype: CD4/CD8= 0,85, CD3 80%, CD4 30%, **CD8 36%**
- Therapy: acetylcystein 3x600mg/daily, Prednisone 1mg/kg/daily, Azathioprine 50mg/daily

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# Sudden respiratory failure

- Peripheral pulse oximetry (PPO): 65%
- Blood gas: pH 7.528 (N:7.35-7.45), pCO2 4,61kPA (N:4.7-6.1), pO2 6,79 kPA (N: 11.04-14.36)
- **Non-invasive ventilation:** PS 15cm, PEEP 6, FiO2 90%, PPO 78%
- **Invasive mechanical ventilation:** pressure-synchronized intermittent mandatory ventilation, inverse I/E ratio, Peak insp pressure 30 and PEEP 10 cm $H_20$, tidal volume 5-8ml/kg, PPO up to 80%, pH 7.318, PO2 **8.02** kPA, pCO2 8.87kPA
- **V-v ECMO:** 3500 rpm, with blood flow from, 3,5 lpm to 4,5 lpm, gas flow through the gas blender 8 lpm with oxygen fraction 1.0. Ventilator settings were: tidal volume 5 ml/kg, oxygen fraction 0.5, pressure plateau, 25 cm $H_2O$, PEEP 10 cm $H_2O$
- Predominantly right heart, multi-organ failure, death

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# Histological pattern

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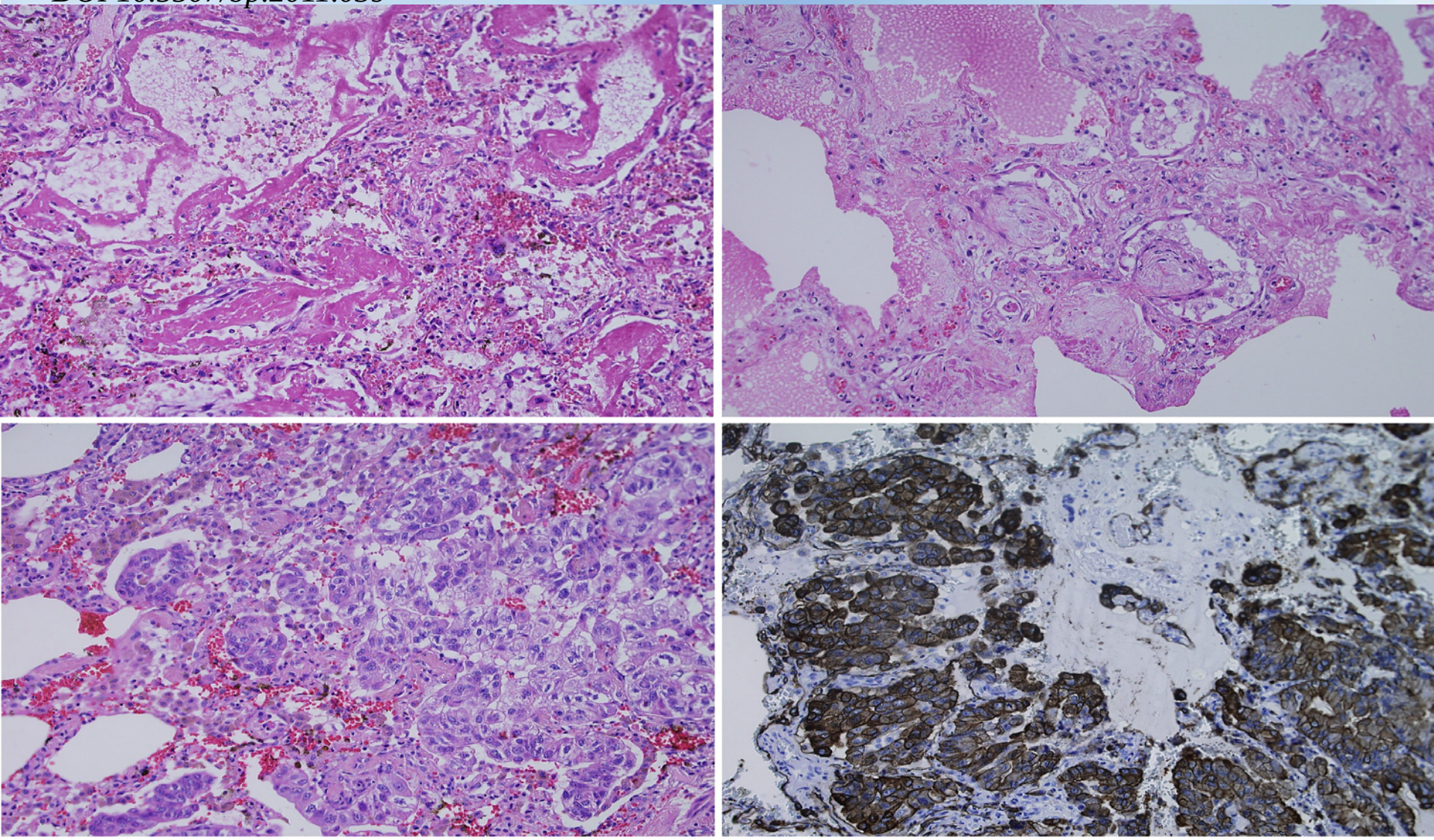

FIGURE 2. Histological demonstration of lung tissue a) with features of diffuse alveolar damage, typical hyaline membranes; part of the alveolar septa with fibroblastic proliferation and patchy alveolar septal infiltrates of mononuclear cells b) with fibroblast foci, usual interstitial pneumonia appearance, spindle-shaped cells in oedematous stroma c) with locus of bronchioalveolar carcinoma d) with locus of bronchioalveolar carcinoma in fibrotic terrain labelled immunohistochemically for cytokeratin 7 as marker of malignant proliferation.
Haematoxylin-eosin staining, 200x magnification, Olympus DP70

# Definitive diagnosis

- **Clinical:** dyspnea > 3 months, fatigue, muscle weakness, respiratory failure
- **Radiological:** dense fibrosis, honeycombing, GGO
- **Histological:** usual interstitial pneumonia+diffuse alveolar damage, + diffuse/multilocular growing bronchoalveolar carcinoma
- **Acute interstitial pneumonia (formerly Hamman-Rich syndrome) in idiopathic pulmonary fibrosis with bronchoalaveolar carcinoma + paraneoplastic myositis**

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# Idiopathic pulmonary fibrosis/Lung cancer

- 67% of IPF patients are at increased risk of lung cancer (LC)[1]
- 21% pacients with LC have had fibrotic interstitial changes at autopsy[2]
- Smoking and male sex are risk factors for LC development in IPF patients[3]
- Confirmed CK and myoglobin elevation in epitelial malignant tumours[4,5,6]


1. Aubry et al. *Mayo Clin Proc.* 2002 Aug;77(8):763-70
2. Meyer EC, Liebow AA. *Cancer.* 1965;18:322-351
3. Park J et al *Eur Respir J*. 2001 Jun;17(6):1216-9
4. Wang HC, Lu JY, Ting YM. Zhonghua Yi Xue Za Zhi 1995;55(3):270-3.
5. Lee BI, Bach PM, Horton JD, Hickey TM, Davis WA. Clin Cardiol. 1985 Apr;8(4):233-6.
6. Flonta SE, Arena S, Pisacane A, Michieli P, Bardelli A. Am J Pathol. 2009;175(1):201-6

# Take home message

- Reduced vital capacity + velcro-like crackles should lead to thorough diagnostic work-up even if x-ray appears normal
- IPF is a risk factor for lung cancer
- Elevation of CK,myoglobin in IPF patients should alert clinicians to probable LC
- Elevation of rheumatological markers may indicate malignant proliferation
- Diffuse growing bronchoalaveolar carcinoma may be hidden under IPF clinical-morphological appearance
- Precaution - imunosupressive therapy in histologicaly unproved dg, where cancer was not excluded

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# Thank you for your attention…


Plasek J, Dvorackova J, Jahoda J, Trulikova K, Mokosova R, Danek T, Hrabovsky V, Martinek A. Acute interstitial pneumonia (Hamman-Rich syndrome) in idiopathic pulmonary fibrosis and bronchoalveolar carcinoma: a case report. Biomed Pap Med Fac Univ Palacky 2011;155(4):289-293 *DOI 10.5507/bp.2011.039*