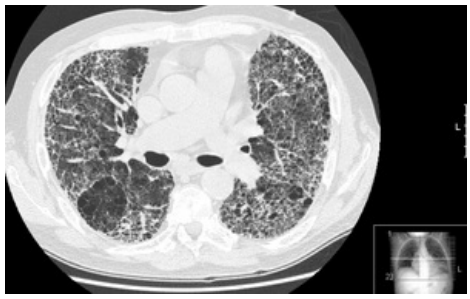
Idiopathic pulmonary fibrosis

It is a diffuse, primarily fibrotic lung process.



HRCT pulmonary fibrosis

Pathogenesis

This is probably a uniform pathological response of lung tissue to both infectious and non-infectious agents. These cause damage to the lining of the alveoli and thus result in progressive and uncontrollable scarring. The inflammatory reaction as such can occur only secondarily.

Epidemiology

- Patients are most often between the ages of 40 and 70.
- The incidence in women is 7.4 / 100,000 and in men 10.7 / 100,000.
- It occurs sporadically, is equally widespread in all localities, familial cases are rare.
- The disease is practically incurable, and even with adequate treatment, survival usually does not exceed 3-5 years.

Clinical picture

- Onset – rolonged unproductive cough in time with worsening exertiona dyspnea, fatigue, weight loss, tachypnoe;
  - on the bases of lungs late inspiratory crepitus similar to **Velcro opening**[1];
  - eventually chronic hypoxia with cyanosis develops.
- In 2/3 of the patients there are club-shaped fingers with nails in the shape of a watch glass.
- Image of COPD without obstructive defect, in the later phase restrictive lung damage - reduction of FVC.
- Despite the typically protracted progressively deteriorating course, acute exacerbations may occur in some patients:
  - sudden clinical deterioration;
  - decreased lung function;
  - radiological image of the so-called milk glass (indicating alveolitis).

Diagnostics

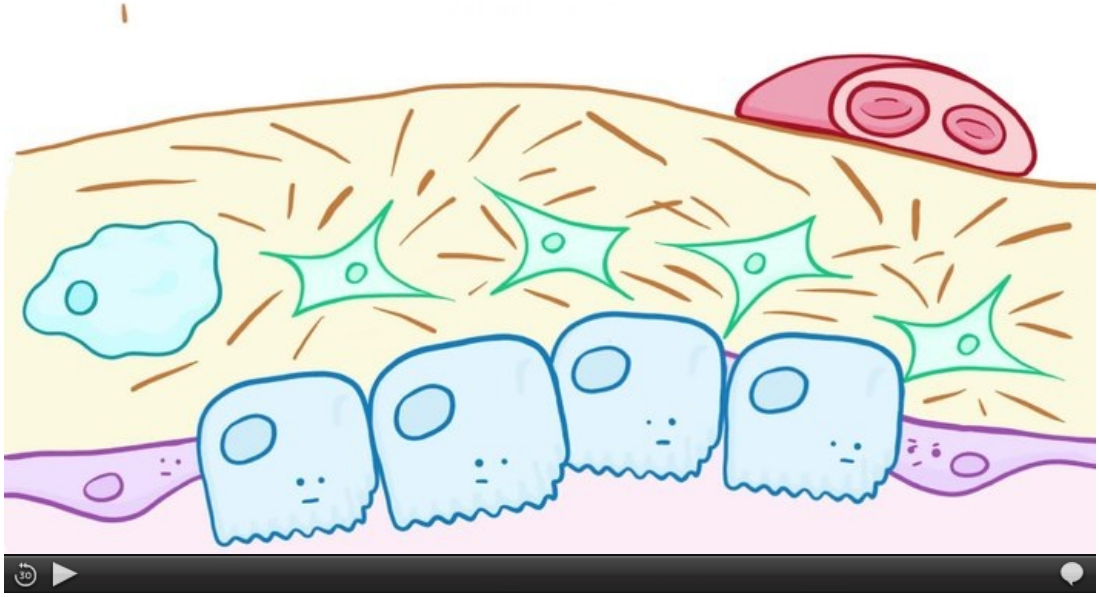
Here, HRCTs are crucial, and a typical clinical finding does not require a biopsy if systemic connective tissue diseases and an exogenous cause are excluded.

- **HRCT image of the lungs**: pulmonary fibrosis with an image of the honeycomb lung in the bases of the lungs and minimal areas of active changes.
- **Histology from a lung biopsy.**
- In patients unable to undergo surgical biopsy, X-ray and bronchoscopy must be sufficient.
- X-ray: increased lung drawing to reticulation - honeycomb lung.
- Functional examination: restrictive ventilation disorder, pulmonary compliance disorder.

Therapy

- Anti-inflammatory and immunosuppressive drugs are ineffective because the main pathological mechanism here is pathological fibroproduction, so they are not used in treatment today.
- Pirfenidon – inhibits fibrosis, indicated in patients with FVC 50-80%. Dosage 3x3cps - a total of 2403 mg.
- Nintedanib – a tyrosine kinase inhibitor on VEGFR, FGFR, and PDGFR
- **Early alveolar lesions**: **N-acetylcystein** 3 times 600 mg (antioxidant effect).
- **Acute exacerbations**: high doses of corticoids, anticoagulant therapy, and antibiotics. **PPI** (proton pump blockers) are given to prevent exacerbations.
- **Advanced diseases with hypoxemia**: long-term home oxygen therapy and consideration of lung transplantation.
- Corticosteroids in long-term therapy **are ineffective**, pbecause fibrotization is not induced by an inflammatory response. [2][3][4]

Summary video



Idiopathic pulmonary fibrosis (video in english)

Links

Related articles

- Chronic lung diseases
- Interstitial lung processes

Reference

1. KOLEKTIV, Marek. *Farmakoterapie vnitřních nemocí - 4. zcela přepracované a doplněné vydání.* - vydání. Grada Publishing a.s., 2010. 777 s. s. 170. ISBN 9788024726397.
2. BENEŠ, Jiří. *Studijní materiály* [online]. [cit. 2010]. <http://jirben.wz.cz>.
3. ČEŠKA, Richard, et al. *Interna.* 1. vydání. Praha : Triton, 2010. 855 s. ISBN 978-80-7387-423-0.
4. VAŠÁKOVÁ, Martina. Léčba idiopatické plicní fibrózy. *Remedia* [online]. 2012, roč. 2012, vol. 2, s. 398-402, dostupné také z <http://www.remedia.cz/Clanky/Farmakoterapie/Lecba-idiopaticke-plicni-fibrozy/6-L-1oN.magarticle.aspx>. ISSN 0862-8947.

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