

CASE FOR DIAGNOSIS

Intrathoracic mass in a 34 year old cystic fibrosis patient

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In 1988, a 29 year old man presented with recurrent purulent bronchitis and sinusitis; cystic fibrosis (CF) was diagnosed. Repeated sweat tests were compatible with the diagnosis of CF: Cl⁻ was 83 (left) and 82 (right) mmol·L⁻¹, and Na⁺ was 95 (left) and 94 (right) mmol·L⁻¹. Chest radiograph at that time was normal. A computed tomography (CT) scan revealed small, tubular bronchiectasis in both upper lobes. Ventilatory function tests showed a mild airway obstruction (forced expiratory volume in one second (FEV₁) 81% predicted, and FEV₁/forced vital capacity (FVC) 66%), with 15% reversibility. Residual volume (RV) was 132% predicted. Lung capacities and CO transfer factor were normal. The patient was atopic, (total immunoglobulin E (IgE) >1,000 kU·L⁻¹; radio-allergosorbent test (RAST) ++++ for *Aspergillus fumigatus*, +++ for *D. pteronyssinus*, ++ for *Cladosporium herbarum*, and + for grass pollen) and precipitation antibodies against *A. fumigatus* were present (two precipitation lines). Skin-prick tests were positive for *D. pteronyssinus* and *D. farinae*, and negative for all other aero-allergens, including *Aspergillus fumigatus*. The patient had azoospermia. Treatment consisted of aerosols (N-acetylcysteine, twice daily), and regular intake of antibiotics, combined with physical therapy in case of purulent bronchitis.

In 1993, the patient was again referred for persistent subpyrexia ($\leq 38^{\circ}\text{C}$), increasing nonproductive cough, anorexia and weight loss (5 kg) of 2 months duration. On examination, the patient was thin (weight 59 kg, height 176 cm), pale, and subpyretic (37.8°C). He had a few slightly enlarged submandibular lymph nodes. There was postnasal drip. Blood pressure was 125/70 mmHg, and pulse rate 100 beats·min⁻¹. Heart sounds were normal. Lung auscultation revealed ronchi over both lung fields.

Erythrocyte sedimentation rate was 86 mm·h⁻¹; and there were 10.9×10^6 cells·L⁻¹ white blood cells (WBC), with 64% neutrophils and 15% eosinophils. C reactive protein was 67 mg·L⁻¹. Pulmonary function tests showed a mild airflow obstruction (FEV₁ 77% pred, FEV₁/FVC 61%); lung volumes (including RV 112% pred) and transfer factor were normal.

Chest radiograph and CT scan of the thorax are shown in figures 1 and 2.

Fibrebronchoscopy yielded an erythematous mucosa in the right upper lobe, bronchus intermedius and orificium of the middle lobe. There was no plugging, but there were some tenacious purulent secretions. Cultures yielded *Staphylococcus aureus*, *Pseudomonas aeruginosa* and *Aspergillus fumigatus*. Mucosal biopsies showed aspecific inflammation; but there were no signs of malignancy.

A parasternal transthoracic tru-cut biopsy was performed (fig. 3)

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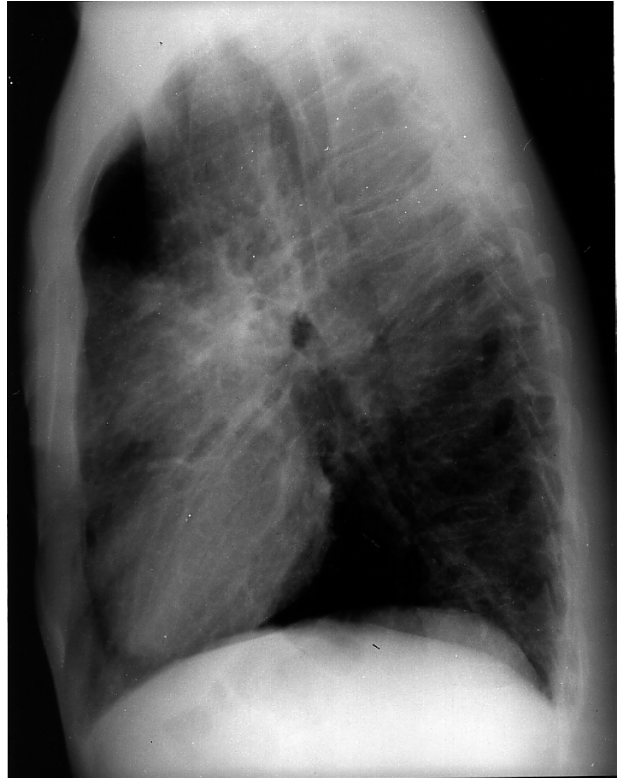
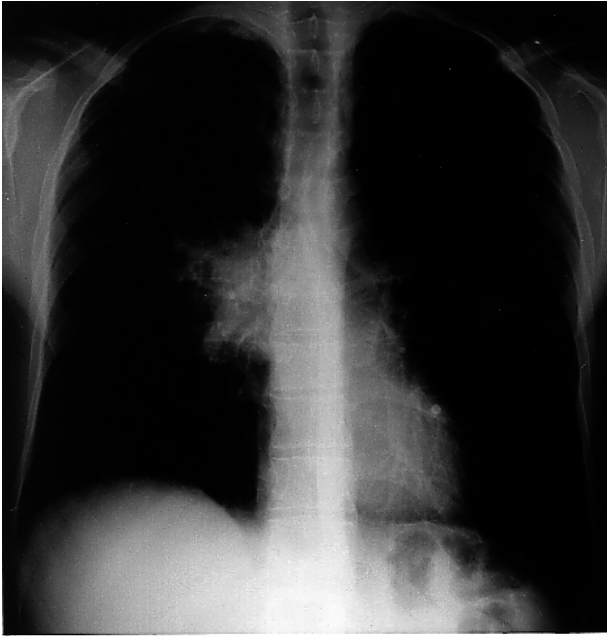


Fig. 1. - a) Posteroanterior (PA) chest radiograph; and b) lateral radiograph.



Fig. 2. - Computed tomography (CT) scan of the thorax.

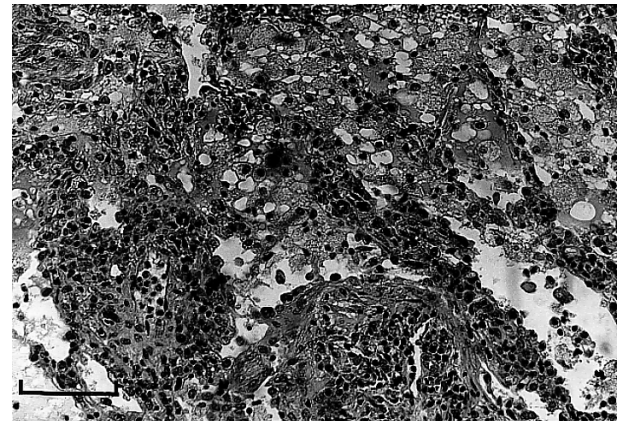


Fig. 3. - Tru-cut biopsy specimen of mass. (Haematoxylin and eosin stain; internal scale bar=100 μ m).

BEFORE TURNING THE PAGE: INTERPRET THE CHEST RADIOGRAPH, CT SCAN AND BIOPSY. SUGGEST A DIAGNOSIS

Interpretation of chest radiograph and CT

The posteroanterior (PA) view (fig. 1a) shows a mass medially in the right hemithorax. The streaks radiating into the lung field, and the left lateral view (fig. 1b), suggest that the mass is situated in the lung, *i.e.* the anterior segment of the right upper lobe.

On the CT scan (fig. 2), however, it is difficult to tell whether the mass is part of the right lung protruding into the left hemithorax, or is located in the anterior mediastinum.

Interpretation of biopsy

Tru-cut biopsies of the mass (fig. 3) consistently yielded cylinders of pulmonary tissue, with a well-preserved architecture of the lung parenchyma. There were focal lesions, characterized by a mixed pattern consisting of thickened septa due to infiltration by nonspecific inflammatory cells, and alveoli filled with foamy macrophages, suggestive of retro-obstructive pneumonia.

A second fibrebronchoscopy performed after the tru-cut biopsy showed patent segmental bronchi in the upper and middle lobe. There were purulent secretions (which consistently yielded *P. aeruginosa*, *S. aureus* and *A. fumigatus*), but there was no visible plugging.

DIAGNOSIS: "Chronic pneumonia of the anterior segment of the right upper lobe, presenting as an anterior mediastinal mass."

Treatment and clinical course

Intravenous antibiotics (ciprofloxacin, netromycin) for 2 weeks, together with aerosolized mucolytics and chest physiotherapy resulted in a complete clinical recovery. Thereafter, aerosolized tobramycin, 80 mg *b.i.d.*, was administered for 6 weeks. Within 2 months, there was a complete disappearance of the "mass" on CT scan (fig. 4).



Fig. 4. - Computed tomography (CT) scan after treatment. The right lung protrudes into the left hemithorax.

Discussion

This patient with CF, diagnosed in middle age, can be situated at the mild end of the clinical spectrum of the disease [1, 2]. Cystic fibrosis - even in its mild form - is a disease of smouldering infection and inflammation, with periodic exacerbations [3]. This chronic pulmonary disease is usually suppurative, most often with infection by *S. aureus*, *P. aeruginosa* and *H. influenzae*. Aspergillus is isolated in about 10% of patients [2-4]. The characteristic feature of CF on chest radiograph [5] is the presence of bronchiectasis, predominantly in the upper zones of the lungs; bronchiolitis (small airway plugging) often results in hyperinflation. Acute infectious exacerbations are typically characterized by zones of focal bronchopneumonia centred around small bronchi and bronchioles. Lobar pneumonia is uncommon in CF, except terminally, and may be associated with necrosis and abscess formation. Lobar or segmental atelectasis is more common, usually in the upper and middle lobes.

This patient presented with a radiographic picture which was initially interpreted as an anterior mediastinal mass; in view of the associated indolent clinical course with subpyrexia, anorexia, malaise and weight loss, lymphoma was at first suspected.

Could this patient also have had allergic bronchopulmonary aspergillosis (ABPA)? He had mild asthma, total IgE was $>1,000$ kU·L⁻¹, specific IgE against *A. fumigatus* was present, there was blood eosinophilia, there were precipitating antibodies against *A. fumigatus*, and he had bronchiectasis and a roentgenographic infiltrate, all criteria for ABPA [6]. This diagnosis was not confirmed, because: 1) the clinical history did not exactly fit the clinical presentation of ABPA; 2) he had no immediate skin reactivity to *A. fumigatus*; 3) the histological specimen was not compatible with ABPA (there were no fungal hyphae in the bronchial lumen, and there were no eosinophils) [6]; and 4) there was a complete recovery with antibiotic treatment without corticosteroids. Using the diagnostic criteria cited above, it is noteworthy, however, that 10% of CF patients also have ABPA [7].

Keywords: Chronic pneumonia, cystic fibrosis.

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