CASE FOR DIAGNOSIS

A patient with ankylosing spondylitis and recurrent haemoptysis

T. Franquet*, N.L. Müller[#], J.D. Flint[†]

Case history

A 54-yr-old male presented with a history of recurrent episodes of haemoptysis over the previous 2 yrs. The patient also complained of back pain and stiffness for 15 yrs due to ankylosing spondylitis. Over the previous 2 weeks, he had been complaining of progressively increasing dyspnoea, malaise and fever. The fever persisted despite antibiotics and the patient showed acute severe haemoptysis. He drank little alcohol and had ceased smoking a few cigarettes daily 20 yrs previously. He had no previous history of pulmonary tuberculosis.

On admission, he was pale and complained of nonproductive cough. His temperature was 37.6°C, pulse rate 98 beats·min⁻¹, respiratory frequency 30 breaths·min⁻¹ and blood pressure 160/90 mmHg. On physical examination, the patient was a thin male, who appeared comfortable at rest, without respiratory stridor. No lymphadenopathy was evident. The head and neck were normal.

Fig. 1.–Posteroanterior chest radiography performed on admission in the upright position.

Fig. 2.–Spiral computed tomography of the chest. Coronal imaging reconstruction.

*Dept of Radiology, Hospital de Sant Pau, Universitat Autònoma de Barcelona, Barcelona, Spain. Depts of [#]Radiology and [†]Pathology, Vancouver Hospital and Health Sciences Centre, University of British Columbia, Vancouver, Canada.

Correspondence: T. Franquet, Dept of Radiology, Hospital de Sant Pau, Avda Sant Antoni M. Claret 167, 08025 Barcelona, Spain. Fax: 34 932 919282. E-mail: 19429tfc@comb.es

Laboratory studies revealed a white cell count of 1.64×10^{10} cells·L⁻¹, haematocrit of 27.3%, platelet count of 4.08×10^{11} cells·L⁻¹ and mean corpuscular volume of 89 μm^3 . The blood sodium concentration was 141 mM, potassium concentration 3.7 mM and chloride concentration 101 mM. Renal and liver function test results were normal. The patient was human immunodeficiency virus-negative. He gave positive human leukocyte antigen (HLA)-27 test results.

Chest radiography (fig. 1) was performed on admission, followed by spiral computed tomography of the chest (fig. 2). On the fifth hospital day, the patient showed severe haemoptysis. Flexible fiberoptic bronchoscopy was performed and revealed minimal secretions and no visible endobronchial lesions. Culture study of a specimen obtained during bronchoscopy was negative for acid-fast bacilli. The results of microbiological culture are shown in figure 3.

Fig. 3.—Photomicrograph of a cytology slide from the aspirated culture (haematoxylin and eosin stain).

BEFORE TURNING THE PAGE, INTERPRET THE RADIOGRAPH, COMPUTED TOMOGRAPHY SCANS, CYTOLOGY RESULTS AND HISTORY AND SUGGEST A DIAGNOSIS AND TREATMENT.

Interpretation

Chest radiography

Posteroanterior chest radiography demonstrated extensive scarring of the upper lobes. A cavitory lesion can be seen in the left upper lung containing an ovoid mass (aspergilloma) and a characteristic crescent of air between the mass and cavity wall. Marked pleural thickening can also be seen surrounding the cavitory lesion. A mildly reticular pattern is evident in the right lung apex.

Spiral computed tomography of the chest

The cross-sectional image demonstrates reticular opacities in the right lung apex. A large cavity containing the aspergilloma is present in the left upper lobe. The coronal reconstruction image demonstrates left upper lobe volume loss, bronchiectasis and communication between the cavity and the left upper lobe bronchus. The fungus ball is separated from the wall of the cavity by an airspace, resulting in the distinctive "air crescent sign" (fig. 2a). The dilated left upper lobe bronchus extending from the cavity to the left hilum is of note. Also of note are reticular opacities in the right lung apex. Significant volume loss from the right upper lobe secondary to lung fibrosis is also visible.

Scoliosis and multiple syndesmophytes (bamboo spine) are visible on the thoracolumbar spine (fig. 2b).

Microbiological study

The microbiological study revealed fungal organisms with broad septate hyphae and 45° branching consistent with *Aspergillus* species.

Diagnosis: "Left apical fibrocavitary disease with saprophytic aspergilloma in a patient with ankylosing spondylitis"

Treatment and clinical course

Embolisation of the left superior intercostal artery was performed. After 5 days, the patient still presented with persistent haemoptysis; repeated embolisation of the left superior intercostal artery was not successful. Owing to the severity of the recurrent haemoptysis, left upper lobectomy was performed.

During thoracotomy, an intracavitary aspergilloma was found in the left upper lobe. Pathological assessment demonstrated a 5×6-cm cavity in the left upper lobe containing large quantities of brown grumous material and communicating directly with a large bronchus, consistent with aspergilloma. Histological sections revealed acute and chronic inflammatory infiltrates in the wall of the cavity. Surrounding the cavity was dense interstitial fibrosis with a severe chronic inflammatory infiltrate, extensive scarring and traction bronchiectasis.

Microscopic examination of the pulmonary specimen revealed an exophytic growth of dichotomously branching septate hyphae compatible with *Aspergillus* species. Culture confirmation of *Aspergillus* species was obtained. During the postoperative period, the patient recovered well.

Discussion

Ankylosing spondylitis is a connective tissue disease of unknown pathogenesis [1–3]. Young males aged 20–40 yrs are the most commonly affected, 90% of whom are HLA-B27-positive [2, 3]. The reported prevalence of pulmonary disease ranges 0–30% [4, 5]. Radiographic manifestations are variable and dependent on the chronicity of the disease [6–9]. ROSENOW *et al.* [4] reviewed the radiographic findings in 2,080 patients with ankylosing spondylitis; 26 (1.3%) of the patients exhibited apical fibrosis or fibrocavitary disease. Although the cause of lung abnormalities remains controversial, bilateral apical scarring is thought to be the earliest pulmonary manifestation of the disease [4]. Bone changes, consisting of symmetric marginal syndesmophytes ("bamboo spine"), are usually visible on the thoracic spine of patients with ankylosing spondylitis who have pleuropulmonary abnormalities. In the series of WOLSON and ROHWEDDER [1], apical fibrosis was found in two (4%) of 52 patients with typical skeletal radiological manifestations of ankylosing spondylitis.

A more extensive spectrum of pulmonary abnormalities has been described using computed tomography. FENLON *et al.* [2] described the radiographic and computed tomography findings in 52 patients with ankylosing spondylitis and pleuropulmonary abnormalities. The most common radiographic findings consisted of interlobular septal thickening, basal interstitial lung disease, bronchiectasis (primary and traction), emphysema, upper lobe fibrosis and pleural thickening.

Cavitation is uncommon. In the series of CROMPTON *et al.* [5], cavitation was found in only one (0.4%) of 225 patients with ankylosing spondylitis. When present, these radiological features may mimic those of end-stage sarcoidosis and chronic tuberculosis with apical fibrocavitary changes. Additional abnormalities such as nonapical fibrosis, bronchiectasis, paraseptal emphysema and tracheobronchomegaly have also been documented [5].

Saprophytic infection with *Aspergillus* (aspergilloma) occurs within a pre-existing pulmonary cavity, often caused by old tuberculosis or sarcoidosis. Aspergilloma has also been described in a small number of patients with ankylosing spondylitis. ROSENOW *et al.* [4] found aspergilloma formation in five (0.2%) of 2,080 cases. Aspergilloma typically leads to conglomeration of intertwined fungal hyphae admixed with mucus and cellular debris within a pre-existent pulmonary cavity or ectatic bronchus [10]. On radiography, aspergillomas are characterised by the presence of a solid round or oval mass with soft-tissue opacity within a lung cavity [10]. Typically, the mass is separated from the wall of the cavity by an airspace of variable size and shape, resulting in the "air crescent" sign [10]. The aspergilloma usually moves when the patient changes position.

Although patients with aspergillomas are usually asymptomatic, they may present with cough and repeated episodes of haemoptysis. Surgical resection remains an option for patients with severe life-threatening haemoptysis [10]. Selective bronchial arterial embolisation may be performed in patients with poor lung function.

References

1. Wolson AH, Rohwedder JJ. Upper lobe fibrosis in ankylosing spondylitis. *Am J Roentgenol Radium Ther Nucl Med* 1975; 124: 466–471.
2. Fenlon HM, Casserley I, Sant SM, Breatnach E. Plain radiographs and thoracic high-resolution CT in patients with ankylosing spondylitis. *AJR Am J Roentgenol* 1997; 168: 1067–1072.

3. Rumancik WM, Firooznia H, Davis MS Jr, *et al*. Fibrobullous disease of the upper lobes: an extraskeletal manifestation of ankylosing spondylitis. *J Comput Tomogr* 1984; 8: 225–229.
4. Rosenow E, Strimlan CV, Muhm JR, Ferguson RH. Pleuropulmonary manifestations of ankylosing spondylitis. *Mayo Clin Proc* 1977; 52: 641–649.
5. Crompton GK, Cameron SJ, Langlands AO. Pulmonary fibrosis, pulmonary tuberculosis, and ankylosing spondylitis. *Br J Dis Chest* 1974; 68: 51–56.
6. Thai D, Ratani RS, Salana S, *et al*. Upper lobe fibrocavitary disease in a patient with back pain and stiffness. *Chest* 2000; 118: 1814–1816.
7. Mayberry JP, Primack SL, Müller NL. Thoracic manifestations of systemic autoimmune diseases: radiographic and high-resolution CT findings. *Radiographics* 2000; 20: 1623–1635.
8. Franquet T. High-resolution CT of lung disease related to collagen vascular disease. *Radiol Clin North Am* 2001; 39: 1171–1187.
9. Geftter WB. The spectrum of pulmonary aspergillosis. *J Thorac Imaging* 1992; 7: 56–74.
10. Franquet T, Müller NL, Giménez A, *et al*. Spectrum of pulmonary aspergillosis: histologic, clinical, and radiographic findings. *RadioGraphics* 2001; 21: 825–837.