A MYCETOMA IN A GENTLEMAN WITH CHURG-STRAUSS SYNDROME

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ABSTRACT

Pulmonary aspergilloma (mycetoma) is a rare pulmonary infectious disease that manifests clinically with fever, chest pain, chronic cough and haemoptysis, although may be asymptomatic in some patients [1].

Here, a case is presented of an elderly gentleman attending hospital coughing up blood-stained sputum. Thoracic computed tomography (CT) revealed a left apical lung cavity with a mycetoma, presumed secondary to an aspergillus infection. The fungal ball dropped anteriorly within the cavity when the patient lay on his front, hence confirming the diagnosis. The haemoptysis settled with Itraconazole; anti-fungals being the treatment-of-choice in symptomatic individuals with a pulmonary aspergilloma.

This gentleman also suffered from severe Churg-Strauss syndrome and required long-term triple immuno-suppressive therapy. Pulmonary aspergillosis is a serious threat to those immuno-compromised as a result of disease or treatment [2]. This case highlights the importance of considering a mycetoma as a cause for respiratory symptoms in immuno-compromised individuals, especially in those presenting with haemoptysis.

CASE PRESENTATION

A 71-year-old male Caucasian ex-smoker presented to hospital with chest pain, exertional dyspnoea and an approximate 3 month history of a cough associated with haemoptysis. It was initially thought that he was suffering from either a lower respiratory tract infection, although he was afebrile on admission and his white cell count and C-reactive protein level were both normal, or congestive cardiac failure. A chest x-ray (CXR) showed a left
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apical lung mass (Figure 1, arrow), following which the differential diagnosis was revised to pulmonary carcinoma, tuberculosis (TB) or a mycetoma. Sputum sent for acid-fast bacilli was negative and a T-spot test was intermediate, thus excluding TB. A subsequent thoracic CT scan was most informative, revealing a large cavity in the left upper lobe measuring approximately 65 x 75 x 85mm that represented a mixed soft tissue density and air containing lesion lying posteriorly within the cavity (Figure 2A, arrow, and 2B) – the appearance being very suggestive of a mycetoma. A repeat CT scan, this time with the patient positioned prone (Figure 2C), demonstrated that the solid component had fallen to the anterior aspect of the cavity (Monad’s sign). The gravity-dependent nature of the radio-opaque fungus ball confirmed the diagnosis. Although serum levels of IgE were not raised and Aspergillus precipitants were negative, symptomatic improvement with Itraconazole, as well as radiological regression of the mycetoma, suggested Aspergillus colonisation as the aetiologcal factor.

Figure 1  
CXR showing a left apical cavitating lesion

This patient suffered from severe Churg-Strauss syndrome with a vasculitic peripheral neuropathy and required long-term immuno-suppressants (prednisolone, cyclosporin and azathioprine). He had no other pre-existing lung disease apart from asthma. This gentleman also reported steroid-induced diabetes mellitus at the time of presentation, as well as a previous left leg deep vein thrombosis (DVT); his admission was subsequently complicated by a Mycobacterium Chelonae skin infection.
Figure 2  Thoracic CT images of the mycetoma

A – coronal view
B – supine transverse view
C – prone transverse view
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DISCUSSION

Pulmonary aspergillosis is a spectrum of mycotic disease that can be subdivided into three main types: saprophytic (most common), allergic and invasive (either chronic necrotising or angio-invasive). It is caused by *Aspergillus* species and, in 80-90% of cases, *Aspergillus fumigatus*: a highly antigenic saprophytic fungus widely distributed in nature and frequently encountered growing on decaying vegetation and damp surfaces [2,3]. This ubiquitous soil fungus is commonly found in the sputum of healthy individuals, although can present as an opportunistic human pathogen that colonises the respiratory tract in susceptible hosts: those individuals with pre-existing lung disease (e.g. bronchiectasis, chronic obstructive lung disease (COPD) and TB) or a depressed immune system [2,3].

Saprophytic aspergillosis, or aspergilloma, is of particular interest in this case, and involves colonisation, without invasion or damage of viable tissue, of a pre-existing cavity, cyst, bulla, or ectatic bronchus in the lung parenchyma. The most common underlying conditions being TB, sarcoidosis, and bronchiectasis [2,3]. This leads to the formation of a fungus ball, or mycetoma, within the cavity, which, histologically, consists of a tangled mass of mycelium, fibrin, inflammatory cells, epithelial-cell debris and other blood products [2,3]. In approximately 50% of the patients, serum precipitin levels are raised [2].

An aspergilloma can present simply as an asymptomatic radiological abnormality in a patient with a pre-existing lung cavity [4]. In fact, about 10% of mycetomas resolve spontaneously [3]. However, haemoptysis is a well-recognised complication, observed in 40-60% of cases [4], which may be substantial and life-threatening [5,6]. In addition, albeit infrequently,
aspergilloma may cause cough and fever [4]. Treatment is instigated only in those patients that develop symptoms, with oral Itraconazole serving as the therapy-of-choice, offering a partial or complete resolution of the mycetoma in 60% of individuals [4]. Several cases also report successful treatment of non-life-threatening symptoms with Itraconazole [7,8,9]. For substantial haemoptysis, surgical resection is curative [1,10], although this is usually performed on patients with an adequate pulmonary reserve [4]. Contraindications to surgery include bilateral advanced lung disease, large transpleural blood vessels, failure to identify the bleeding site, and continued haemoptysis after previous surgery [3]. In patients with intractable or recurrent haemoptysis who are not fit for surgery, bronchial artery embolisation is an effective recourse [11,12], as is percutaneous CT-guided intra-cavity injection with amphotericin paste [13,14].

OUTCOME AND FOLLOW-UP

This patient’s mycetoma responded well to Itraconazole in terms of symptomatic and radiological improvement. Nonetheless, he was readmitted several months later with severe bilateral pneumonia, developed another left leg DVT, venous infarction of the right foot (phlegmasia cerulea dolens), acute renal and liver failure and subsequently died.

LEARNING POINTS

Globally, 15-20% of patients with lung cavities at least 2cm in diameter will develop an aspergilloma [3]. It is important to include this as part of the differential diagnosis in an immuno-suppressed individual complaining of respiratory problems, particularly haemoptysis. If a patient presents in such a way, prompt treatment with Itraconazole should be commenced to avoid severe and fatal blood-loss.

REFERENCES