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A Rare Case of Bilateral Aspergillomas in a Patient of Ankylosing Spondylitis

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ABSTRACT

Pulmonary involvement by *Aspergillus sp.* mainly includes allergic bronchopulmonary aspergillosis, aspergilloma, and invasive aspergillosis. Aspergilloma (Fungal ball) is the most common form of aspergillous pulmonary involvement, which occurs in preexisting pulmonary cavities, especially secondary to pulmonary tuberculosis. Ankylosing spondylitis is a rare cause of upper lobe fibro-cavitary lesions in pulmonary parenchyma. It may also lead to development of fungal balls in pulmonary cavities. Most common presentation is mild to massive hemoptysis; dyspnoea, chronic cough, expectoration may be other presentation; even the patient may remain asymptomatic. Intaracavitary mobile mass is a valuable sign for fungal ball, best detected by computed tomography (CT) scan of thorax. Lobectomy is the treatment of choice to stop the hemoptysis, if the general condition of the patient is fit; otherwise associated co-morbidities complicate the post-operative scenario. In this situation, bronchial artery embolization may be used as a temporary measure to control hemoptysis. Here, we report a case of bilateral aspergillomas within the cavities located in upper lobes of both lungs in a 74 years old male who was suffering from ankylosing spondylitis for last 42 years.

KEY WORDS

Aspergilloma, ankylosing spondylitis, hemoptysis, lobectomy

INTRODUCTION

The fungus *Aspergillus sp.* very rarely gives rise to human infection. Spectrum of aspergillus involvement of pulmonary parenchyma varies from allergic bronchopulmonary aspergillosis (in asthmatics), allergic alveolitis (in non-asthmatics), saprophytic involvement like aspergilloma or fungal balls (in pre-existing lung cavities) to invasive or disseminated aspergillosis in immunocompromised, debilitated patients, associated with various co-morbidities (opportunistic infection).[1] Among different species, *Aspergillus fumigatus* is the most common offending agent.[2] Open-healed tuberculous cavity is most common preceding lesion of aspergilloma.[3] Other causes of pulmonary parenchymal fibro-cavitary lesions in upper lobes are allergic bronchopulmonary aspergillosis, chronic sarcoidosis, chronic hypersensitivity pneumonitis, cystic fibrosis (CF), non-CF bronchiectasis, silicosis, ankylosing spondylitis, eosinophilic granuloma, talcosis, neurofibromatosis, radiation etc.[4] Thoracic involvement of ankylosing spondylitis includes apical fibrosis (with or without cavitary lesions), fungal balls, interstitial lung disease, emphysema, bronchiectasis, pleural thickening, spontaneous pneumothorax, calcifications and ossifications of paraspinal ligaments, stiffness of costovertebral and costosternal joints, stiffness of thoracic spine etc.[5,6] Pleuro-parenchymal involvement in ankylosing spondylitis is uncommon, and bilateral pulmonary aspergillomas occur occasionally. Here, we report a case of bilateral aspergillomas in upper lobes of both lungs in a 74 years old male, suffering from ankylosing spondylitis.

CASE REPORTS

A 74 years old normotensive, non-diabetic, non-smoker male teacher presented to us with gradually progressive shortness of breathlessness of insidious onset for last seven years with cough and scanty, white, mucoid expectoration for same duration. During last six months, he developed five episodes of massive hemoptysis. Last two episodes required hospitalization and blood transfusion. Last episode was nine days back which was followed by ongoing streaky hemoptysis, not responding to conservative medical management. At presentation, the patient was breathless at rest, but there was no orthopnoea or paroxysmal nocturnal dyspnea, and postural, diurnal or seasonal variation. He also gave the history of three episodes of exacerbation of shortness of breath and cough with expectoration of moderate amount, yellow purulent sputum and high grade fever during last seven years, which were controlled by different antibiotics. There was no chest pain, wheeze, fever, and leg swelling. No history of malaise, anorexia, fatigue, and weight loss was documented. There was history of progressively increasing low back pain and stiffness, pain with swelling of wrist, ankle, and hip joints for last 42 years. His neck movement was restricted. He was diagnosed as having ankylosing spondylitis on the basis HLA B27 positivity and radiological evidence of sacroiliitis from outside. Antinuclear antibody (ANA), anti-ds DNA auto-antibody, anti-CCP antibody, and ANCAs were negative with normal serum uric acid level. He received irregular treatment for ankylosing spondylitis comprising of steroids, non-steroidal anti-inflammatory drugs, and methotrexate during last 40 years, but the disease severity was progressively increasing. He denied giving a history of suffering from pulmonary tuberculosis of intake of anti-tubercular drugs. Repeated sputum examination for acid fast bacilli were done for respiratory ailments, but they were always negative, but radiological evidence of progressively increasing upper zone fibrosis in lung parenchyma was noted on serial chest radiographs.

On general survey, anemia and clubbing were noted, but no cyanosis, edema, lymphadenopathy, and engorged neck veins were seen. His temperature was 97°F, respiratory rate-26 breaths/minute, pulse rate-102 beats/minute, and blood pressure-110/60 mmHg. Examination of respiratory system revealed bilateral restricted movement of chest wall with infra-clavicular hollowness, more on the right side. Trachea was shifted to right side and apex beat was not localized. Intercostal suction was present and accessory muscles of respiration were working. Expansion of the chest was 3 cm. Rib crowding was present on both sides, more at right axillary area. Vocal fremitus was increased on right infra-clavicular area. Percussion note was impaired on both sides along mid-clavicular line from second intercostal space downwards up to fourth intercostal space on right side and third intercostal space on left side. Diminished vesicular breath sound was heard over left infra-clavicular area, and cavernous bronchial breath sound was noted over right infra-clavicular area with increased vocal resonance over the same area. Coarse crackles were audible over both infra-clavicular and axillary areas. Examination of musculoskeletal system revealed rigidity of thoracic spine with kyphosis and restricted movement of cervical spine, atlanto-axial and atlanto-occipital joints. No swelling and tenderness in the joints of the four limbs was noted. The Patrick's test in the left leg revealed pain on contralateral side posteriorly due to sacroiliitis. The Schober's test was positive, indicating reduced flexion of lumbar spine. Examination of other system including eye revealed no abnormality.

Complete hemogram revealed only anemia and raised erythrocyte sedimentation rate (56 mm) and blood biochemistry was normal. Sputum for acid fast bacilli was negative and its mycobacterial culture showed no growth after 6 weeks. Gram stain was negative and pyogenic culture showed no growth. Chest X-ray (postero-anterior view) showed bilateral fibro-cavitary lesions with fungal balls and bamboo spine appearance of thoracic region. Contrast enhanced computed tomography revealed similar findings and change of position of intra-cavitary mass with change of patient's posture was noted. (Fig. 1)

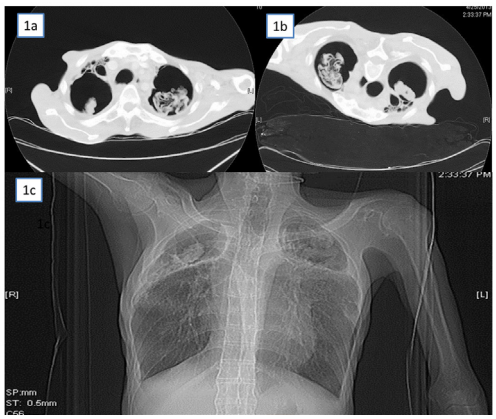

Figure 1. CECT thorax showing bilateral fungal balls within the cavities (1a) with change of position with change of posture (1b) and bamboo spine appearance (1c).

Spirometry showed restrictive defect. Sputum for fugal stain was negative. Anti-HIV – 1 and 2 antibodies were non-reactive. Serum immunoglobulin G level against Aspergillus fumigates increased. Fiberoptic bronchoscopy revealed no endobronchial lesion and analysis of bronchoalveolar lavage fluid taken from upper lobes of both sides showed fungal elements in smear and growth of aspergillus on fungal culture. Ultrasonography of abdomen revealed no abnormality. Fine needle aspiration cytology of right sided intra-cavitary mass lesion revealed only inflammatory cells and no evidence of tuberculosis or malignancy was documented. HLA B27 was again positive, although ANA, anti-ds DNA auto-antibody, anti-CCP antibody, and rheumatoid arthritis factor were still negative. Hence the diagnosis was bilateral fibro-cavitary lesions in upper lobes with fungal balls, complicated by massive hemoptysis.

Conservative medical management failed to stop the bleeding. Right upper lobectomy was done, but the patient died on first post-operative day due to type I respiratory failure. On histopathology of the resected specimen showed septate filamentous fungal hyphae (Fig. 2) and fungal culture revealed the growth of *Aspergillus fumigatus*. So finally the diagnosis was bilateral aspergillomas within the fibro-cavitary lesions of upper lobes, which was secondary to ankylosing spondylitis.

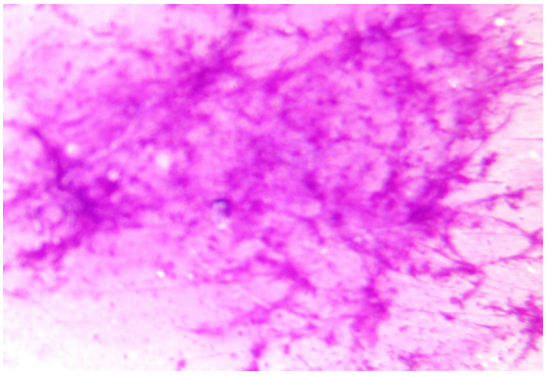

Figure 2. Microphotograph of histopathology of upper lobectomy specimen showing filamentous fungal hyphae with inflammatory cells (H&E stain, 10x).

DISCUSSION

Bilateral upper lobe pulmonary parenchymal fibrosis is most commonly due to pulmonary tuberculosis in our country which belongs to tuberculosis endemic zone. Aspergilloma or fungal ball is defined as a mass consisting of fungal elements, inflammatory cells, mucus, fibrin, and tissue debris formed in the pre-existing lung cavity.[2] Aspergilloma within the fibro-cavitary lesions of the upper lobe is a common complication, but concurrent bilateral involvement is rare, especially in ankylosing spondylitis where occurrence of bilateral upper lobe lung fibrosis is unusual.

In case of ankylosing spondylitis, initially there are apical consolidations which are followed by gross fibrosis, mainly in the upper zones, and sometimes may extend to the mid zones.[7] It starts with unilateral involvement, later becomes bilateral. The incidence of apical fibrosis in ankylosing spondylitis is low, ranging from 1.3–30% and associated with prolonged course of the disease.[8] Histologically, initially there is patchy pneumonia with round cell and fibroblastic infiltration followed by interstitial fibrosis, later dense fibrosis and bronchial dilatation which usually progresses, with the formation of bullae and cavities.[9] The proposed causes of the fibrosis are recurrent aspiration leading to aspiration pneumonitis, alterations in apical mechanical stress from a rigid thoracic spine, and recurrent impaired cough secondary to alterations in respiratory mechanics.[9,10] The clinical presentation and radiological appearance usually simulate to chronic tuberculosis, but there is no evidence that it is caused by tuberculosis, other bacterial infections, or by radiation. It occurs as an extra-articular manifestation of ankylosing spondylitis.[5,8] These cavities may be colonized by saprophytic fungi like aspergillus to form fungus balls or mycetomas. Initially inhalation of airborne spores of Aspergillus gets them deposited deep to the damaged lung parenchyma where they grow on the walls of the cavities due to inadequate drainage and form aspergilloma.[2]

The patients are usually presenting with shortness of breath, cough, and expectoration due to predisposition to recurrent infections, and hemoptysis, which may be scanty or massive. Recurrent infections in fibrotic lung, mycelial micro-invasion of blood vessels on the cavity wall, fungal endotoxins with hemolytic properties, and mechanical friction of the fungal ball with the blood vessels on the cavity wall are the proposed causes of bleeding.[2] Shortness of breath is progressive due to disease progression and caused by pulmonary parenchymal fibrosis and rigidity of thoracic skeleton due calcification and ossification of joints' ligaments and bone involvement due to ankylosing spondylitis itself. Lung function test reveals restrictive pattern. In a small number of cases, the patients are asymptomatic.[11]

Massive hemoptysis is a life threatening condition and mainly due to aspergillomas in pulmonary fibro-cavitary lesions.[12] It usually needs lobectomy as all efforts with the conservative medical management are in vain.[13] Bronchial artery embolization may be used as a temporary measure, and recurrence is inevitable.[14] Postoperative complications, even death is common, as baseline lung function is grossly compromised due to the disease itself. Medical management of ankylosing spondylitis with immunosuppressants and biological (steroids, methotrexate, infliximab, etanercept etc.) should be given to halt the disease progression and therefore its complications. The medical treatment of ankylosing spondylitis predisposes recurrent pulmonary infections and tuberculosis which should be considered as an etiology of bilateral upper lobe fibrosis.

In our case, bilateral upper lobe fibro-cavitary disease due to irregularly treated ankylosing spondylitis was complicated by bilateral aspergillomas which was unusual and rare. The patient presented to us massive hemoptysis due to aspergillomas and died from type I respiratory failure in first post-operative day following right upper lobectomy, probably due to severely compromised baseline lung reserve. Inadequate management of underlying disease probably resulted in clinico-radiological progression of bilateral upper lobe fibrosis without any evidence of pulmonary tuberculosis in recent and remote past and fibro-cavitary lesion of lung parenchyma ultimately leads to development of fungal balls in both sides simultaneously – an uncommon occurrence.

CONCLUSION

Bilateral fibro-cavitary lesions in upper lobes of lungs occur most commonly in pulmonary tuberculosis as a sequelae, especially in our country, India, whch is situated TB endemic zone. But these lesions rarely occur in ankylosing spondylitis which is further complicated by bilateral aspergillomas. These fungal balls are presented by massive haemoptysis which is the dreadest complication of aspergilloma. Surgical removal is the only treatment to control this life threatening haemoptysis permanently.

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