OLGU SUNUMU CASE REPORT

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ASPERGILLOMA AND IDIOPATHIC PULMONARY FIBROSIS: A RARE COEXISTENCE

ASPERGİLLOMA VE İDİYOPATİK PULMONER FİBROZİS: NADİR BİRLİKTELİK

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Öz

Pulmoner aspergilloma (miçetoma) genellikle önceki tüberküloz, fibrotik sarkoidoz veya interstisyel akciğer hastalıkları dahil fibrokaviter hastalıklar nedeniyle gelişen kistik ve kaviter lezyonlara kolonize olan Aspergillus türlerine bağlı gelişen saprofitik bir fungal enfeksiyondur. İdiyopatik pulmoner fibrozis (İPF), nadir, kronik ilerleyici fibrotik akciğer hastalığıdır ve olağan interstisyel pnömoni histolojisi ile karakterizedir. Diyabetik ve ağır sigara içicisi olan 69 yaşındaki erkek hasta kuru öksürük, nefes darlığı, ateş ve kilo kaybı şikayetleri ile kliniğimize başvurdu. Pulmoner aspergilloma ve İPF birlikteliği tanısı yüksek çözünürlüklü bilgisayarlı tomografi bulgularına göre kondu. Burada, İPF zemininde Aspergillus enfeksiyonu gelişmesi ile ilgili olan bu nadir olguyu literatür eşliğinde sunmak istedik.

Anahtar Kelimeler: Aspergilloma, İdiyopatik pulmoner fibrozis, Fungus topu, Kavite

Abstract

Pulmonary aspergilloma (mycetoma) usually is a saprophytic fungal infection due to aspergillus species which is colonized and grows into pre-existing cystic/cavitary lesions resulting from pulmonary tuberculosis or fibrotic sarcoidosis, or other fibro-cavitary diseases including interstitial lung diseases. Idiopathic pulmonary fibrosis (IPF) is a rare, chronic, progressive fibrotic lung disease, characterized by the histological pattern of usual interstitial pneumonia. Heavy smoker, diabetic, a 69-year-old male patient was admitted to our clinic with complaints of dry cough, fever, dyspnea on exertion and weight loss. The diagnosis was made as coexisting of pulmonary aspergilloma and IPF according to findings on HRCT. Hereby, we wanted to present this rare case regarding of coexisting of the development of Aspergillus infection and idiopathic pulmonary fibrosis in the light of the literature.

Keywords: Aspergilloma, Cavity, Fungus ball, Idiopathic pulmonary fibrosis

Introduction

The form in pulmonary involvement of aspergillus infection depends on the hosts' immune status and the local lung structure. Pulmonary Aspergillus infections are usually classified as pulmonary aspergilloma, allergic bronchopulmonary aspergillosis (ABPA), chronic necrotizing pulmonary aspergillosis (CNPA), and invasive aspergillosis [1]. Endobronchial aspergilloma, the other unusual form, is characterized by the growth of the Aspergillus species into the bronchial lumen with or without a parenchymal lesion and/or

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cavity and sometimes may be associated with CNPA or ABPA [2,3].

Pulmonary aspergilloma (mycetoma) (fungus ball) is a saprophytic fungal infection due to aspergillus species that are colonized and grows into pre-existing cystic/cavitary lesions or enlarged airways. Although it frequently develops in a preexisting lung cavity resulting from pulmonary tuberculosis or fibrotic sarcoidosis, it has been reported in a variety of other fibro-cavitary diseases such as bronchiectasis, lung abscess, echinococcal cyst, granulomatous polyangiitis, tumors, and interstitial lung diseases including idiopathic pulmonary fibrosis (IPF) [1,4-7].

IPF is a chronic, irreversible and progressive fibrotic lung disease with unknown etiology, characterized by the histological pattern of usual interstitial pneumonia.[8] The worldwide incidence is 10.7/100.000 person-years for men and 7.4/100.000 person-years for women. Its prevalence is estimated to be 20/100,000 persons for males and 13/100,000 persons for females.[9] IPF has a poor prognosis with median survival times of between 2-5 years.[9]

To the best of our knowledge, only one case has been reported about the coexistence of idiopathic pulmonary fibrosis and aspergilloma [1]. This rare coexistence is presented here as the second time.

Case Report

A 69-year-old male patient was admitted to our clinic with complaints of dry cough, fever, dyspnea on exertion, and weight loss for 9 months. He had a diagnosis of diabetes mellitus and hyperthyroidism. He had no history of past pulmonary tuberculosis or known active contact, no exposure to environmental pollutants/dust, no gastroesophageal reflux disease, but smoked 150 packed-year cigarettes. He also had an operation (right thoracotomy+wedge resection) from the right lower lobe due to pulmonary aspergilloma three years ago, but no clue for pulmonary interstitial fibrosis on the pathology report. After the operation, he received Voriconazole treatment for one year. On admission, his general condition was good, conscious, and cooperative. His blood pressure was 100/60 mmHg, heart rate was 86/min, and respiratory rate was 14/min. On auscultation, Velcro rales were heard on the posterobasal lung fields bilaterally. All the other systemic examinations were normal, including no finger clubbing. In laboratory, Hb: 12,8 g/dl, WBC:7000/mm3, Hct: 39%, ESR: 8mm/h, C-reactive protein: 23mg/L, and routine biochemical analysis were in normal limits, including thyroid function tests. Urine analyses were also normal. Acid-fast bacilli smears and cultures of the sputum were also found negative for tuberculosis. His oxygen saturation (Sat O2) was 91% at rest. He failed to complete the 6-minute walking test, walked only 97 meters, and was desaturated (79%). His arterial blood gas analysis showed pH: 7.47, pO2: 80.2 mmHg, pCO2: 32.7 mmHg, HCO3: 23.5 mmol on the room air. Pulmonary function tests revealed a moderate restrictive pulmonary dysfunction. He was unable to comply with the diffusion capacity for the carbon monoxide (DLCO) test. Chest X-ray revealed increased peripheral reticular densities bilaterally, and a cavitary lesion at the right upper zone (figure 1).

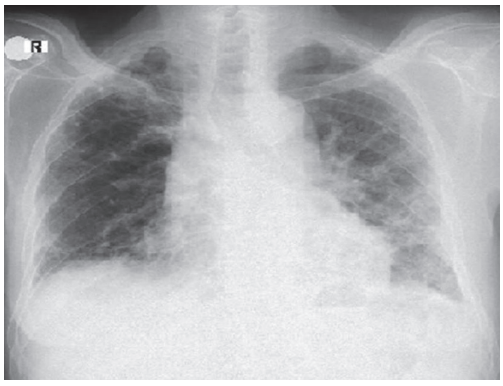

Figure 1
Postero-anterior Chest X-ray of the patient.
Chest X-ray of the patient showing a suspected cavitary lesion and diffuse bilaterally reticular shadowing and evident volume loss.

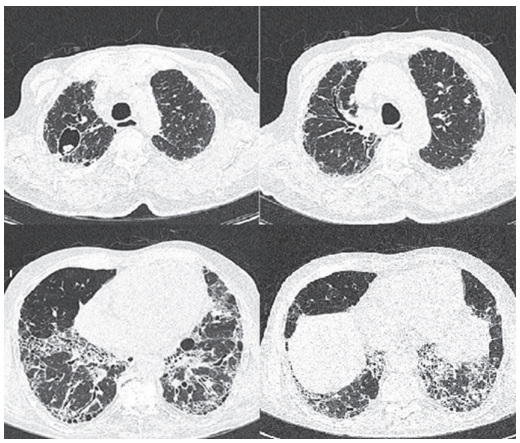

Figure 2
High-Resolution Computerized Tomography sections of the patient.
HRCT sections showing a fungus ball within a cavitary lesion at the right upper lobe, and tractional bronchiectasis, interlobar/interlobular septal thickening and honeycombing images at the other lung areas bilaterally.

Thorax CT showed increased subpleural interlobular/interlobar septal thickening, tractional bronchiectasis, and honeycombs in lower lobes and also a cavity (22x30mm) with a fungal ball in the right upper lobe (figure 2). Echocardiography showed no evidence for pulmonary hypertension. Evaluation with a fiberoptic bronchoscopy revealed a normal appearance of the tracheobronchial tree, inconclusive bronchoalveolar lavage (BAL), and no microbiologic proofs for pulmonary tuberculosis or aspergillosis. Anti-nuclear antibodies, anti-dsDNA, rheumatoid factor, anti-neutrophil cytoplasmic antibodies, and other rheumatologic tests were negative. The specific IgE value for the aspergillus fumigatus was 0.33kUL (0-0.35). Since the patient did not accept any diagnostic or therapeutic surgical intervention, both the Voriconazole and the Pirfenidone treatments were started. For symptomatic purposes, long-acting B2 agonist and anticholinergic inhaler treatments besides long-term oxygen therapy were also given.

Discussion

Pulmonary aspergillosis may occur in different ways depending on the underlying lung structure and host immune status.[10] Aspergilloma is a fungal ball consisting of dead and living mycelial elements, fungal fibrin, amorphous debris, mucus, inflammatory cells, and degenerating blood and epithelial elements. Most aspergillomas are caused by *A. fumigatus*, but rarely by *A. niger* especially in diabetics and/or active tuberculosis.

In immunocompetent patients, the colonization of aspergillus species to the bronchial lumen requires structural changes that induce airflow stasis. In our case, he had no history of pulmonary tuberculosis, but his chest X-ray and HRCT showed destructive, fibrotic changes of the parenchyma that resulted in airway narrowing/enlarging and obstruction.

The diagnosis of aspergilloma depends on clinical, radiological, and serological/microbiological pieces of evidence. The most common symptoms of the disease are cough, sputum, and hemoptysis, although a patient with aspergilloma can remain asymptomatic for years. Massive/fatal hemoptysis is the commonest indication for surgery. The important clue in the diagnosis of aspergilloma is its unique image on radiology. Fungal ball movement within the cavity can often be shown radiologically with the change of body position if the ball does not fill the entire cavity.[11] The presence of air-crescent signs in HRCT findings suggests a high incidence of aspergilloma. In our case, a fungus ball inside a cavity, very suggestive for aspergilloma, was detected on the HRCT scan. But sometimes, aspergilloma may mimic or coexist with a lung tumor. [12] Pulmonary aspergilloma inside of a lung cavity can be visualized by bronchoscopy in some cases of endobronchial aspergilloma.[2] In our case, bronchoscopic examination showed that the tracheobronchial tree was normal, and BAL culture was negative for aspergilloma, although it may be positive in more than half of patients. Precipitating antibodies to Aspergillus antigens are present in the sera of >95% of patients with aspergilloma.[1] Other diagnostic procedures such as transthoracic fine-needle aspiration or surgical removal of the lesion may be considered with keeping the potential risks in mind, as we did in our case.

CT has a central role both in detecting early changes and smaller lesions of aspergillomas and making definitive/probable IPF diagnoses without the need for a lung biopsy. In the early stages, CT may depict the initial fungal fronds arising from the cavity wall, which subsequently detaches and coalesce to form the classic intracavitary mass with an adjacent air crescent. [13]

In pulmonary aspergilloma, the most common and the most effective treatment is surgical resection, which is usually reserved for patients who have massive hemoptysis with favorable pulmonary reserve. Voriconazole has limited therapeutic effects on this type of aspergillus infection. In our case, chronic symptoms related to IPF were controlled with pirfenidone, but radiological dimensions of the cavity and the fungus ball remained stable after four months of therapy with Voriconazole.

In immunocompetent patients, aspergilloma may develop within the preexisting honeycomb structures and traction bronchiectasis due to excessive fibrotic processes which are generally seen on the chronic basis of IPF which is actually not a cavitary disease.

Kaynaklar

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