

## Central Bronchiectasis with Allergic Broncho-pulmonary Aspergillosis- A Case Report and Review of the Literature

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### ABSTRACT

Allergic bronchopulmonary aspergillosis is an allergic immune response to colonization of the airways with *Aspergillus fumigatus*. It is a complex disease characterised by cough with mucus production in the form of characteristic plugs and bronchiectasis. Here we report a case of Allergic bronchopulmonary aspergillosis in a 58 year old female patient and review its clinical, radiological, cytological features and treatment aspects.

**Keywords:** *Aspergillus*, Allergic bronchopulmonary aspergillosis, Asthma, Bronchiectasis, Cystic fibrosis.

### Introduction

Patients with bronchiectasis show permanent dilatation of bronchi, which is due to the damage of bronchial and bronchiolar walls due to inflammation and infectious processes.[1] Allergic bronchopulmonary aspergillosis (ABPA) is a pulmonary disorder due to a hypersensitivity reaction to persistent *Aspergillus fumigatus* in the airways.[2]

ABPA is usually seen in patients suffering from asthma or cystic fibrosis (CF), particularly those associated with atopy. Patients complain of symptoms that are due to their primary disease. Management is directed against the allergic inflammatory response. Early diagnosis and treatment is necessary to prevent progression of disease, parenchymal damage and loss of lung function.[3] ABPA has a complex pathophysiology. In an allergic person, the persistence of *A. Fumigatus* in the lung leads to T lymphocyte activation and immunoglobulin (Ig), cytokine release and inflammatory cell infiltration. This results in mucus production, airway hyper reactivity and eventually bronchiectasis.[4]

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Patients complain of cough with sputum and wheezing is seen. Sputum typically is in the form of tan to brownish-black mucus plugs. Mucus plugs consists of mucin, degenerating eosinophils and desquamated epithelial cells. Main features of ABPA are bronchiectasis, mucus plugging, lobar collapse and fibrosis. Central bronchiectasis in multiple lobes is suggestive of ABPA. Whereas, central bronchiectasis is less severe and generally limited to one or two lobes in patients with asthma.[5, 6]

Hallmark of ABPA is that the total serum IgE levels of minimum 1,000 IU/ml. Patients show elevated levels of *A. fumigatus* specific IgE, IgG antibodies, precipitins and eosinophils.[2] Chest radiographs reveal fleeting parenchymal opacities or bronchiectasis. Infiltrates are usually eosinophilic in nature and at times wrongly diagnosed as infectious pneumonia.[7]

The main aim of management is to prevent additional lung destruction by suppressing the inflammatory pathway so that does not occur. A number of medications have been tried in the treatment of ABPA. These comprise systemic and inhaled corticosteroids, antifungal agents and omalizumab, a monoclonal antibody directed against IgE. [2, 8]

### Case Report

A 68 year old female patient came to the department of pulmonology, Prathima Institute of Medical Sciences,

Karimnagar, Telangana State, with complaints of cough with expectoration, shortness of breath and fever since 10 days. The cough was insidious in onset. It was mucoid initially, mucopurulent later. Sputum was of moderate quantity, non foul smelling. There was shortness of breath, which was sudden in onset, gradually progressed to Grade III-IV, and was associated with wheeze. Fever was insidious in onset, low grade, continuous and not associated with chills. She gave a history of similar complaints last year, which subsided with medication. There is no history of

hypertension, diabetes, bronchial Asthma, epilepsy and tuberculosis.

**Examination:** Examination of the patient revealed that the patient was conscious, coherent, cooperative, moderately built, well nourished. Vital signs were BP : 120/80 mm, PR : 106/min, Temperature was 98.4°F. SpO2 – 92% on room air. Lungs examination showed decreased breath sounds in right infra-scapular and infra-axillary areas, bilateral rhonchi present. Patients coughed up well-formed, tan to brownish-black mucus plugs (Fig 1)



Fig 1: Mucous plugs

Differential diagnosis of Right lower lobe collapse with Pleural effusion, Right Pleuro-parenchymal fibrosis and Right lower lobe mass with Pleural effusion was given.

**Investigations:** CECT chest revealed bilateral central bronchiectasis and mucous plugging of the right

bronchus intermedius causing collapse of the right lower and middle lobes. Collapsed lung showed contrast enhancement with crowding of the dilated bronchioles (Fig 2 and 3). Hence bronchoscopic intervention might save the function of the collapsed lobes.



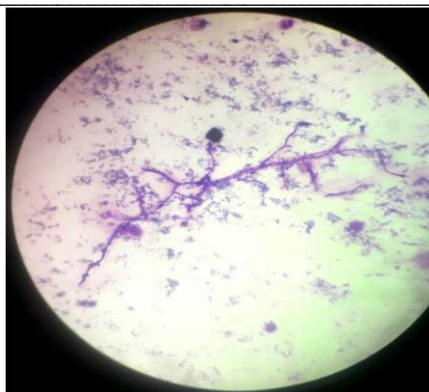
Fig 2: CECT chest -Coronal view



Fig 3: CECT chest -Sagittal view

Fibro-Optic Bronchoscopy revealed copious amount of thick mucopurulent secretion in right upper lobe, intermediate and lower lobe bronchi. Cytological

examination revealed multiple aggregates of eosinophils and Charcot-Leyden crystals present in a background of mucus. Several fungal hyphae with features of *Aspergillus* were also seen (Fig 4).



**Fig 4: Cytology**

A final diagnosis of Central Bronchiectasis with Allergic Broncho-pulmonary Aspergillosis was given.

## Discussion

*Aspergillus* is a fungus that is seen all over the world and grows optimally at body temperature. Spores are small and easily aerosolized and get deposited in distal and terminal airways. ABPA is an idiopathic pulmonary disease, characterized by an allergic inflammatory response to colonization of the airways by *A. fumigatus*. [2, 8, 9]

ABPA can exist in two diverse forms [5, 6]

1. ABPA-seropositive (S): Patients have a history of asthma, investigations show total IgE >1000 IU/mL, elevated serum anti- *A. fumigatus* IgE and IgG, positive immediate hypersensitivity skin test to *A. fumigatus*; and/or serum anti-*A. fumigatus* IgG antibodies.

2. ABPA-central bronchiectasis (CB): Along with the criteria of ABPA-S, characteristic findings of advanced disease like expectoration of mucus plugs, sputum culture positive for *A. fumigatus* and central bronchiectasis.

Diagnosis of ABPA is made by clinical, radiographic and serologic features. A positive an immediate IgE-mediated response skin test and IgG-mediated late skin test response is variably positive in ABPA. A wheal of minimum 3 mm diameter, elevated total IgE, *A. fumigatus* specific antibody levels assist to differentiate ABPA from other conditions like asthma with *Aspergillus* sensitivity. [2, 10] The differential diagnosis for ABPA comprise of refractory asthma, newly diagnosed cystic fibrosis, tuberculosis, sarcoidosis, infectious pneumonia, eosinophilic pneumonia and *Aspergillus* sensitive asthma. [2, 11] Rosenberg, Patterson (1977) suggested a set of diagnostic criteria for ABPA. They included primary and secondary criteria. Primary criteria include episodic bronchial obstruction, peripheral eosinophilia, positive immediate skin test to *Aspergillus*, positive precipitin

test to *Aspergillus*, increased total serum IgE, history of transient or fixed lung infiltrates and proximal bronchiectasis. If patients have 1-6 criteria, it is suggestive of ABPA and if all 7 criteria are met, then ABPA is definite. Secondary or supportive criteria include brown plugs/flecks in sputum, positive late (6–12 hours/Arthus) skin test to *Aspergillus*. [2, 11]

Later on with advancements in the field of diagnostic medicine, the diagnostic criteria for ABPA have been modified in the form of Modified ISHAM working group 2013 criteria for diagnosis of ABPA. They include predisposing asthma or cystic fibrosis, obligatory criteria like IgE > 1000 IU/mL and positive immediate skin test or increased IgE antibody to *Aspergillus*, supportive criteria include eosinophilia > 500, precipitins or increased IgG antibody to *Aspergillus* and CECT chest 1) central bronchiectasis 2) Finger in glove appearance. [2, 12] Treatment consist of systemic corticosteroids as they are the mainstay of therapy for ABPA. Inhaled corticosteroids, oral antifungal agents, standard airway clearance treatments. Mucus clearance with Ambroxol, bromhexine, Guanaphensin, N-acetyl cysteine etc, chest physiotherapy and therapeutic bronchoscopy are suggested. [2, 13-15]

## Conclusion

The fungal genus *Aspergillus* is everywhere in the environment and hence *Aspergillus* spores inhalation is inevitable. ABPA is a complex hypersensitivity response to *A. fumigatus*. Even now the underlying pathogenesis of ABPA is still not understood properly and it is evolving. We suggest that one has to understand the clinical features of ABPA, so as to

diagnose and treat the disease earlier. In present case we suggest that early detection and radiological diagnosis with the tools like Contrast enhanced CT test and early interventional bronchoscopy save the above patients lung function from permanent disability and improved the quality of life. We suggest that one has to understand the clinical features of ABPA so as to diagnose and to treat the disease earlier.

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