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# The Ghost in the Closet. Allergic Sino-Bronchopulmonary Aspergillosis Without Bronchial Asthma: A Case Report & Review of the Subject

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**SUMMARY**

Allergic bronchopulmonary aspergillosis (ABPA) is a complex hypersensitivity reaction, often in patients with asthma or cystic fibrosis (CF), which occurs when there is colonization of airways by Aspergillus species. In the absence of bronchial asthma, the diagnosis of ABPA is questionable but the reported patient had no features of bronchial asthma and presented with multifocal radiological shadows in both the lung fields which on further exploration revealed obscure ABPA.

## INTRODUCTION

Allergic bronchopulmonary aspergillosis causes repeated episodes of bronchial obstruction, inflammation, and mucoid impaction which can lead to permanent damage in the lungs including bronchiectasis, fibrosis, and respiratory compromise.

The clinical picture of ABPA is dominated by asthma complicated by recurrent episodes of bronchial obstruction, expectoration of brownish mucus plugs, peripheral blood and pulmonary eosinophilia, and occasionally hemoptysis. There is no individual test to establish the diagnosis of ABPA and the condition is confirmed by utilizing various clinical, radiographic, and immunologic criteria. Treatment of ABPA aims to control episodes of acute inflammation and to limit progressive lung injury. Glucocorticoids are most commonly used, although there is evidence of benefit from combined therapy with itraconazole. Early detection and treatment may reduce the risk of progression to fibrotic disease.

## CASE REPORT

A 24 years non smoker ranger's police soldier, resident of Burewala presented with long standing history of chronic persistent allergic rhinitis. A month ago, he underwent surgery in ENT unit because of bilateral nasal obstruction due to nasal polyps & fungal balls extensively involving nose and paranasal sinuses on both sides, with extension into the right orbit violating lamina papyracea but without intracranial extension. Prior to his ENT surgery, he had taken complete two courses of anti tuberculous treatment on the basis of cough & mucoid sputum production for the last 3 years along with abnormal chest radiographic findings. Along with other investigations, he also had his p-ANCA and c-ANCA in the suspicion of necrotizing granulomatous vasculitis (former Wagener's granulomatosis) but both were negative. He was referred to the pulmonology OPD for bronchoscopy in the suspicion of drug resistant tuberculosis.

On further questioning, he denied any respiratory symptoms except cough with mucoid

sputum during episodes of rhinorrhoea & post nasal dripping for which he was using antihistaminic drugs. There were no other triggers for cough & there was no history of any wheezing during or prior to this illness and no nocturnal awakening due to shortness of breath. On evaluation, he was an average built young man with a pulse of 76/m, blood pressure 110/70 mmHg and afebrile with a respiratory rate of 18. Chest examination was normal except a few occasional crackles bilaterally. Other systems examination was also normal. Bedside SpO2 was 97% on air & PEFR 650L/m.

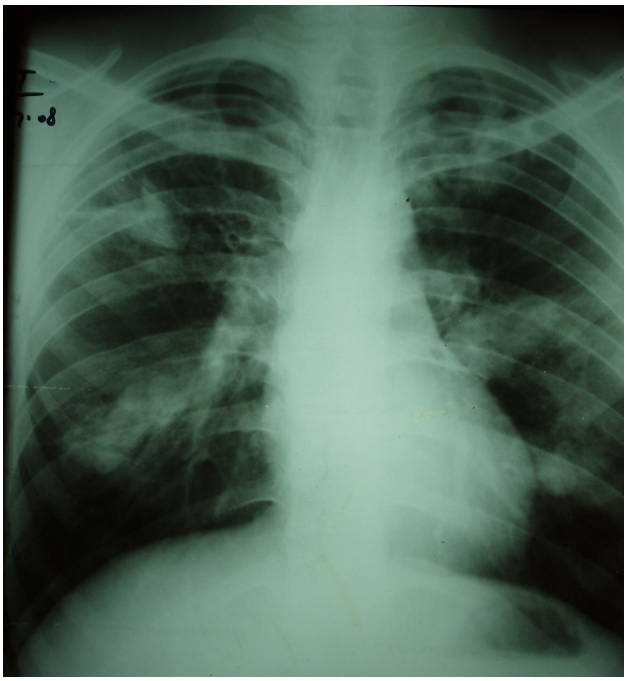

**Fig. 1: CXR-PA: Irregular & inhomogeneous patches and typical gloved finger shadows.**

His chest radiograph showed bilateral irregular & inhomogeneous patches and typical gloved finger shadows (Fig. 1). HRCT chest showed bilateral bronchiectatic changes involving upper lobes along with cystic areas having air fluid levels and patchy opacities in both lungs suggestive of mucoid impaction (Figs. 2 and 3). Radiologist had suggested the possibility of fungal infection versus tuberculosis.

His spirometry showed normal shape of flow volume loop and all maneuvers within the normal range including $FEV_1$, FVC, $FEV_1$/FVC and

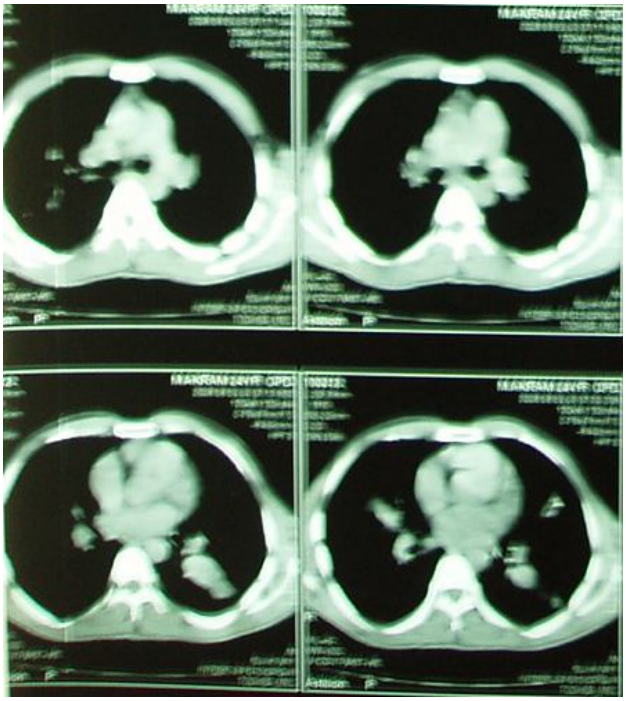

**Fig. 2: HRCT chest showing bilateral shadowing suggestive of mucoid impaction.**

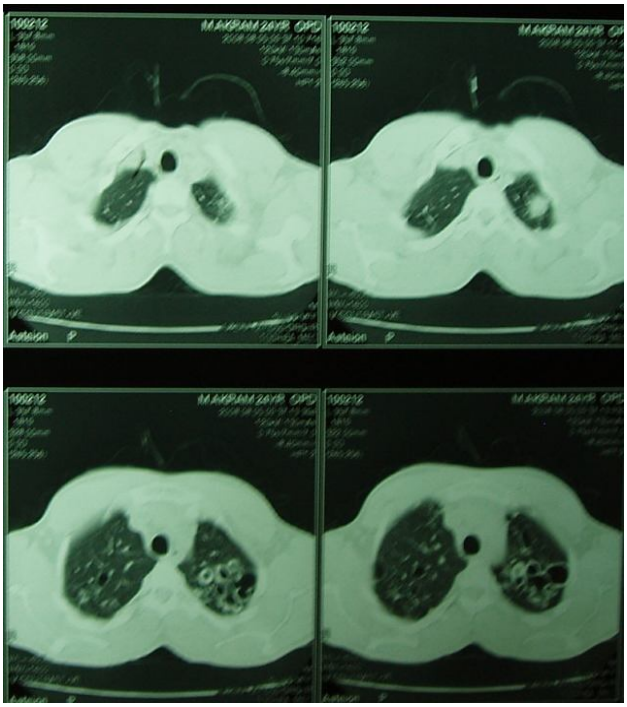

**Fig. 3: HRCT chest showing bilateral bronchiectatic changes involving upper lobes.**

FEF 2575 without any significant reversibility post bronchodialatation (Fig. 4). Although his spirometry and clinical findings contradicted the presence of bronchial asthma which is the top criterion for ABPA diagnosis, the provisional diagnosis of ABPA was still considered on the basis of typical chest x-rays findings as described above and HRCT showing bronchiectasis (one of the major criteria). The diagnosis was confirmed when further investigations revealed: Positive skin prick testing for aspergillus, raised serum IgE level to 8300u/L, peripheral blood eosinophil level of 14%, positive anti aspergillus IgG (titre 15 u/ml) and bronchial washings showed budding aspergillus hyphae while negative mycobacterium staining and culture.

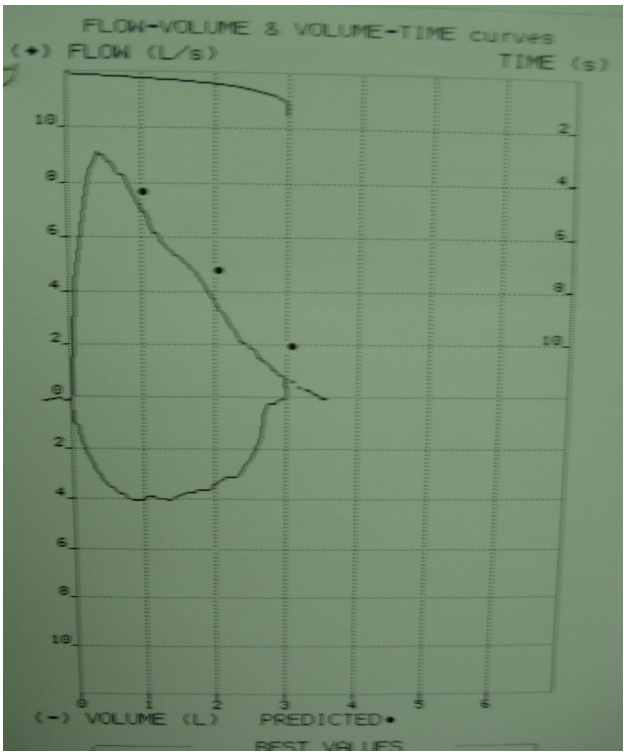


**Fig. 4: Spirometry: Normal shape of flow volume loop except earlier termination of exhalation**

He was treated with a tapering regimen of prednisolone for 6 months and itraconazole for 16 weeks. He showed marked clinical and immunological improvement (serum IgE 1350u/L) with considerable positive changes in the chest radiograph (Fig. 5).

## DISCUSSION

Members of the genus Aspergillus are ubiquitous in nature and therefore everyone has daily contact with their spores, and total avoidance is virtually impossible. These saprophytic molds exist wherever organic matter is decomposing and thrive on fallen leaves and compost heaps. More than 100 species have been identified, but over 95% of human illness is caused by Aspergillus fumigatus.[1] The spectrum of human illness is extensive, ranging from allergic reactions to colonization of preexisting pulmonary cavities to invasion and destruction of lung tissue with pyemic spread to brain, skin, and other organs and rapid death.[2]

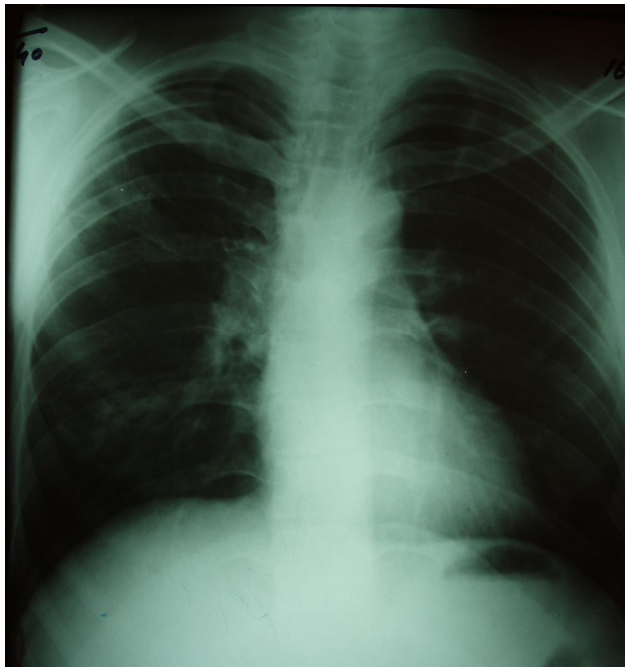

**Fig. 5: Post-treatment chest radiograph is almost clear of patchy shadowing**

Allergic fungal sinusitis (AFS) has been described as the upper airway equivalent of allergic bronchopulmonary aspergillosis. The clinical presentation is that of recurrent or chronic sinusitis with nasal polyposis, which is often refractory to prolonged antibiotic therapy.[3] Thick, tenacious mucus, called allergic mucin, is often present in surgically obtained specimens. With appropriate

staining, infiltrating eosinophils, lymphocytes, and fungal hyphae can be demonstrated in this material.[4]

Pathologically ABPA is characterized by mucoid impaction of the bronchi, eosinophilic pneumonia, and bronchocentric granulomatosis in addition to the histologic features of asthma.[1] Septated hyphae with acute dichotomous branching may be seen in the mucus-filled bronchial lumen, but fungi do not invade the mucosa (only colonize).[5] Radiologically there are parenchymal infiltrates (usually involving the upper lobes), atelectasis due to mucoid impaction, and a number of findings characteristic of bronchiectasis including tram line shadows, ring shadows, toothpaste shadows and gloved finger shadows due to intrabronchial exudates with bronchial wall thickening.[6] Compared to a chest radiograph, high resolution computed tomography (HRCT) scan of the thorax may show widespread proximal cylindrical bronchiectasis with upper lobe predominance and bronchial wall thickening.[7]

The prime criterion is the presence of bronchial asthma among the major diagnostic criteria for classic ABPA but there are reports of patients presenting without clinical asthma.[8,9,10] During a retrospective survey of patients with pulmonary shadows and blood eosinophilia, 42 patients were found with allergic bronchopulmonary fungal disease and 11 of these had no clinical evidence of asthma. In the absence of asthma there was some difficulty in making a diagnosis, particularly where collapse of the upper lobe occurred in middle-aged or elderly patients and bronchogenic carcinoma was presumed responsible.[10] It is therefore suggested the diagnosis be considered in patients with lung disease and blood eosinophilia even in the absence of asthma. Others diagnostic criteria for ABPA include: Positive skin prick test to aspergillus, precipitating serum antibodies to A. fumigatus, serum total IgE concentration >1000 ng/mL, peripheral blood eosinophilia >500/$mm^3$, lung infiltrates on chest x-ray or chest HRCT, central bronchiectasis on chest CT and elevated specific serum IgE and IgG to A. fumigatus.[11] Treatment of ABPA aims to control episodes of acute inflammation and to limit progressive lung injury with utilization of corticosteroids to be the primary agents.[12] There is also recommendation of addition of itraconazole for sixteen weeks in all patients who require substantial doses of glucocorticoids, with the goal of enabling a reduction in the glucocorticoid dose.[13]

## REFERRENCES


1. Stevens DA, Moss RB, Kurup VP, Knutsen AP, Greenberger P, Judson MA et al. Allergic bronchopulmonary aspergillosis in cystic fibrosis- State of the art: Cystic Fibrosis Foundation Consensus Conference. Clin Infect Dis. 2003; 37:225-64.
2. Ben-Ami R, Lewis RE, Kontoyiannis DP. Enemy of the immunosuppressed state: an update on the pathogenesis of Aspergillus fumigatus infection. Br J Haematol 2010; 150:406.
3. Hutcheson PS, Schubert MS, Slavin RG. Distinctions between allergic fungal rhinosinusitis and chronic rhinosinusitis. Am J Rhinol Allergy. 2010; 24: 405.
4. Hamilos DL. Allergic fungal rhinitis and rhinosinusitis. Proc Am Thorac Soc. 2010; 7: 245.
5. Riscili BP, Wood KL. Noninvasive pulmonary Aspergillus infections. Clin Chest Med. 2009; 30: 315.
6. Buckingham SJ, Hansell DM. Aspergillus in the lung: diverse and coincident forms. Eur Radiol. 2003; 13: 1786.
7. Johkoh T, Müller NL, Akira M, Ichikado K, Suga M, Ando M et al. Eosinophilic lung diseases: diagnostic accuracy of thin-section CT in 111 patients. Radiology. 2000;216:773.
8. Kornfeld H, Mark J. A 34-Year-Old Woman with One Cystic Lesion in Each Lung. N. Engl. J. Med. 1999; 341: 974-82.
9. Beer DJ, Mark J. A 57-Year-Old Man with a Chronic Productive Cough, Dyspnea, and Extensive Bilateral Air-Space Disease. N. Engl. J. Med.1994; 330:1599-1606.
10. JJ Glancy, Elder JL, McAleer R. Allergic bronchopulmonary fungal disease without clinical asthma. Thorax. 1981. 345-34.
11. Greenberger PA. Allergic bronchopulmonary

aspergillosis. J Allergy Clin Immunol. 2002;110:685.

12. Agarwal R, Gupta D, Aggarwal AN, Saxena AK, Saikia B, Chakrabarti A et al. Clinical significance of decline in serum IgE levels in allergic bronchopulmonary aspergillosis. Respir Med. 2010;104:204.
13. Salez F, Brichet A, Desurmont S, Grosbois JM, Wallaert B, Tonnel AB. Effects of itraconazole therapy in allergic bronchopulmonary aspergillosis. Chest. 1999;116:1665.

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