Allergic Bronchopulmonary Aspergillosis: An interesting presentation

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Abstracts

Allergic Bronchopulmonary Aspergillosis (ABPA) is an under diagnosed respiratory condition, which very often masquerades as Bronchial Asthma and results in a delay in identification and subsequent treatment. It is one of the components of the spectrum of aspergillosis – which is a group of diseases caused by the fungi Aspergillus fumigatus. It occurs due to an exaggerated response of the immune system to the fungi. It has a wide variety of presentations, most commonly occurring as fleeting opacities on chest x rays. We present an interesting case of ABPA having a unique presentation.

Keywords: consolidation, bronchiectasis, aspergillosis.

1. Introduction

Allergic bronchopulmonary aspergillosis (ABPA) is an indolent and progressive disease, resulting from a hypersensitivity response in the airways to Aspergillus fumigatus.[1] This fungus is responsible for a variety of respiratory conditions, commonly referred to as spectrum of aspergillosis. Host characteristics are a major determinant of the type of pulmonary disease in response to aspergillus exposure.[2] ABPA was first reported by Hinson and his colleagues[3], followed by a detailed description by Pepys and coworkers.[4,5] It commonly occurs in patients with Bronchial asthma and cystic fibrosis, the precise incidence of which is not known. The most important clue to a diagnosis of ABPA is bronchial asthma not responding to usual treatment. While a diagnosis of ABPA is difficult, its treatment is directed at mitigating the allergic inflammatory response.[6]

2. Case report

A 42 year old female was brought to our outpatient department by onlookers in a semiconscious state. Her pulse was feeble, blood pressure was 100/60 mm Hg and she had a saturation of 80%. Her extremities were cold and clammy and she was cyanosed. On auscultation she had tubular bronchial breath sounds all over the right hemithorax. A history of the patient could not be obtained, as she was brought in a semiconscious state.

She was immediately admitted to the ward. Her chest x ray on admission showed a homogenous opacity in the right hemithorax, with deviation of the trachea towards the right, and a suspicious looking cystic lesion in the right mid zone. A provisional diagnosis of right sided fibrocystic collapse with post tuberculous COPD was made and the patient was managed with nebulised bronchodilators, supplemental oxygenation, intravenous steroids and antibiotics.

To our surprise, the patient responded within a day, and subsequently did not need supplemental oxygen. She denied a past history of tuberculosis on repeated questioning. She underwent a high-resolution computed tomography on the fourth day of admission, which showed bilateral areas of central bronchiectasis, more on the right, with complete disappearance of the homogenous opacity observed on the chest x ray. A repeat chest x ray also showed bilateral bronchiectasis.

Her serum IgE was 2432 IU/ml and her specific IgE against Aspergillus fumigatus was negative. This patient was hence diagnosed as a case of ABPA – central bronchiectatic variant. Her spirometry demonstrated aforced vital capacity (FVC) of 82%, forced expiratory volume in 1st second (FEV1) of 62% with reversibility of 240 ml and 16% and a FEV1/FVC ratio of 68. She was hence
started on inhaled bronchodilators and 1 mg/kg of oral prednisolone, to be subsequently tapered off. This patient continues to follow up regularly in our outpatient department.

Figure 1: Chest x ray on admission, showing right sided fibrocystic collapse

Figure 2: High resolution computed tomography of thorax showing bilateral central bronchiectasis with complete disappearance of the homogenous opacity seen in the x ray at admission

Figure 3: Chest x ray showing bilateral areas of bronchiectasis, right more than left

3. Discussion

Allergic bronchopulmonary aspergillosis (ABPA) is an inflammatory pulmonary syndrome complicating the course of various pulmonary disorders, mainly bronchial asthma and cystic fibrosis (CF).[7] It is the most widely studied Aspergillus-related allergic phenomenon, is an immune-mediated inflammatory syndrome caused by hypersensitivity to a ubiquitous fungus, Aspergillus fumigatus.[8] On the other hand, allergic bronchopulmonary mycosis (ABPM) is an ABPA-like syndrome due to fungal organisms other than A. fumigatus. The frequency of ABPM is negligible compared to that of ABPA.[9]

The causative agent is ubiquitous in the environment, hence it is easy for susceptible people inhale their spores regularly, leading to a constant exposure to an antigenic stimulus. Genetic susceptibility to the agent plays a very important role in the pathogenesis of ABPA. Aspergillus fumigatus is unique, as an aeroallergen, and inhalation alone is insufficient to lead to ABPA. Persistence of the viable fungus within the airways seems to be an important factor in determining the development of ABPA. Viable Aspergillus has been found growing on and between bronchial epithelial cells, despite an intense inflammatory cell infiltrate,[10] whereas its proteases lead to the release of pro-inflammatory mediators from epithelial cells.[11,12] This constant inflammatory process, if not halted leads to permanent damage of the pulmonary architecture and eventually leads to end stage lung disease.

While the natural course of ABPA is difficult to predict, a strong clinical suspicion is of utmost importance. In local settings, any patient who is having “difficult to treat” asthma, must be evaluated for the presence of ABPA. In a country like India, the diagnosis of cystic fibrosis is a rarity. Evaluation must compulsorily include serological evaluation like serum IgE levels as well as radiological investigations including high resolution computed tomography of the thorax.

As mentioned before, the clinical progression of ABPA is hard to predict, and majority of the patients suffer due to delay in diagnosis. ABPA, clinically, goes through 5 stages – acute, remission, exacerbation, steroid dependent and fibrotic.[13,14] The diagnosis is usually done in either the acute or the exacerbation stage. The main aim of the treating physician is to maintain the patient in the remission phase.

There is no simple test that can establish the diagnosis of ABPA and a set of criteria is required for this purpose. Greenberger laid down eight diagnostic criteria in 1997.[15] However, there is still no consensus on the number of criteria used to
diagnose ABPA. ABPA has been classified by Patterson et al on the basis of HRCT chest findings as ABPA-CB and ABPA-S, depending on the presence or absence of bronchiectasis.[16] It was hypothesized by Greenberger et al. that ABPA-S is the earliest stage of ABPA, with less severe immunologic findings than in ABPA-CB.[17] Even though this is the most widely accepted classification of the types of ABPA, there have also been other attempts at classification. Kumar et al subsequently divided ABPA into three groups, namely, ABPA-S, ABPA-CB, and ABPA-CB-ORF, the last group being ABPA with central bronchiectasis and other radiological features, including fibrosis, bullae, blebs, pleural thickening etc.[18] Agarwal et al included a new subtype known as ABPA-HAM (highly attenuated mucous).[19]

The best thing about ABPA is its response to oral corticosteroids, which further necessitates an earlier diagnosis. Oral corticosteroids suppress both the immune response as well as the inflammation. Different studies and reviews have recommended different dosages and duration of therapy.[20,21] The authors prefer starting oral corticosteroids at the dose of 1 mg/kg for 6 weeks and then gradually tapering it by 5 mg every week, for a minimum duration of 6 months. The duration of treatment varies from case to case and may also extend upto 1 year. Controversy surrounds the usage of anti fungal agents in ABPA. Itraconazole has been tried at the dosage of 200 mg twice a day for 6 months, with different results. However, the authors firmly believe that long term trials are a must before any recommendation can be made.

Thus our patient, who presented to us in exacerbation, was wrongly diagnosed as right sided fibrocystic collapse and post tuberculous COPD. It was only after a detailed evaluation, that she was labelled as a case of ABPA – CB variant.

4. Conclusion

ABPA is a complex hypersensitivity response to A. fumigatus in atopic patients with bronchial asthma. It is characterized by worsening symptoms, serum eosinophilia, and fleeting pulmonary opacities on radiographs. An elevated serum total IgE, central bronchiectasis, and hyper attenuating mucus on chest CT scan heightens the suspicion for ABPA. An early diagnosis is required if permanent damage to the pulmonary architecture has to be arrested. Early identification allows treatment with corticosteroids with a potential role for newer antifungal azole medications as corticosteroid-sparing agents, which may improve the long-term outcome in this potentially relapsing condition.

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