WHEEZING AND CYSTIC FIBROSIS CHILDREN

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Abstract

Cystic fibrosis is the most frequent monogenic disease in population with Caucasian origin, potentially lethal, with marked clinical variability. Wheezing is a common symptom in cystic fibrosis, asthma, allergic bronchopulmonary aspergillosi , even in tuberculosis . Sometimes these pathological conditions are difficult if not impossible to be differentiated. The aim of the paper was to evaluate the frequency of co morbidities like asthma, ABPA or TB in children with CF. Methods: One hundred and twenty-four children with CF were evaluated. For the retrospective analysis on the frequency of associated TB, asthma, ABPA, we used data records from our CF Centre. Results: A small percent of these children 7.25% were diagnosed with associated asthma. Thirteen children (10.4%) were diagnosed with sensitization to ABPA, 3 of them had aspergillosis, with rapid decline of lung function. Regarding the tuberculosis, only one patient (<1%) with CF had criteria for TB diagnosis. Interestingly, before being diagnosed with CF, almost 13% of patients were considered and treated as TB cases, most of them with predominant respiratory symptoms. Conclusion: ABPA is significant co morbidity in CF patients, while asthma occurs rarely. Although TB is a quit common condition in our area, CF children seemed to be protected against it. Further studies need to be done to evaluate this hypothesis.

Key words: wheezing, cystic fibrosis, children

Background

Cystic fibrosis (CF) is the most frequent monogenic disease in population with Caucasian origin, potentially lethal, with marked clinical variability. Wheezing is a common symptom in asthma, allergic bronchopulmonary aspergillosis (ABPA), cystic fibrosis (CF), even in tuberculosis (TB). Sometimes these pathological conditions are difficult if not impossible to be differentiated.

The aim of the paper was to evaluate the frequency of co morbidities like asthma, ABPA or TB in children with CF.

Methods

One hundred and twenty-four children with CF , aged between 5-21 years, with median age at diagnosis = 11.34 years were evaluated. Study design: observational, retrospective for ten years period. For the retrospective analysis on the frequency of associated TB, asthma, ABPA, we used data records of CF Centre.

Diagnosis criteria for ABPA used IN the study were according European Cystic Fibrosis Society:

*Two of three criteria:*Immediate skin reactivity to *Af* antigen, Precipitating antibodies to *Af* antigen, Total serum IgE . 1,000 IU/mL and at least two of the following: Bronchoconstriction, Peripheral blood eosinophils. 1,000/mL, History of pulmonary infiltrates, Elevated serum IgE/IgG specific to *Af*, *Af* in sputum by smear or culture *Af* , Response to steroids.

The diagnosis of asthma was suggested by the following: episodes of acute airway obstruction reversed by bronchodilators(especially if seasonal), a strong family history of asthma and/or evidence of atopy, or laboratory evidence of allergy such as eosinophilia or elevated IgE; This definition is reasonable although the use of serum IgE and eosinophilia is only of value if allergic bronchopulmonary aspergillosis (ABPA) has been excluded. Briefly, diagnosis of asthma require: history of atopy+FEV1 variation >20% +positive bronchodilation test +/-increase IgE and eosinophilia; positive skin prick test or IgE specific For diagnosis of tuberculosis, positive Mantoux test , BAAR positive smear and bacteriological confirmation, additional to clinical and radiological signs were considered. As investigation, spirometry tests, radiography or CT scan were used when necessary.

Results

A small percent of these children 7.25% were diagnosed with associated asthma. Thirteen children (10.4%) were diagnosed with sensitization to ABPA, 3 of them had aspergilosis (fig. 1), with rapid decline of lung function. (ABPA) is a lung disease caused by an immunologic response to the mold *Aspergillus s*pp.

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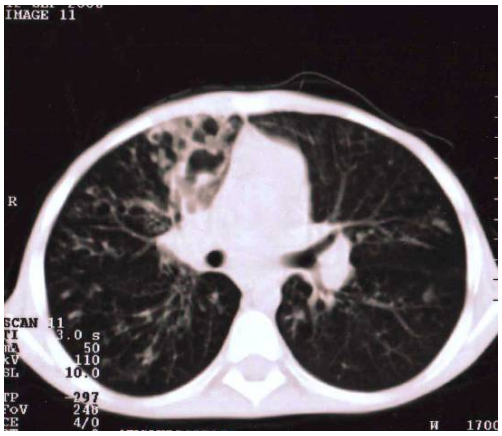

Fig.1. A case of aspergillosis in bronchiectasis in CF child.

Regarding the tuberculosis, only one patient (<1%) with CF had criteria for TB diagnosis (fig. 2). Interestingly, before being diagnosed with CF, almost 13% of patients were considered and treated as TB cases, most of them with predominant respiratory symptoms.

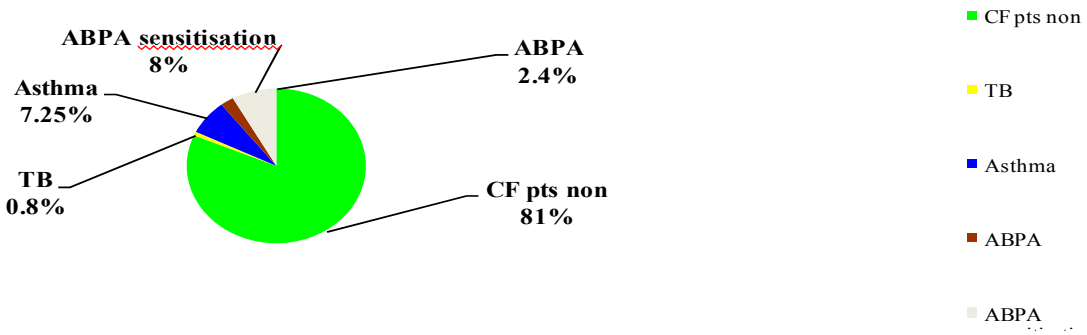

Fig. 2. Distribution of wheezing disorders in CF children.

Estimates and descriptions of the prevalence of ABPA and of sensitization to A. fumigatus based only on classic criteria, even in developed countries, may be underestimated, since the difficulties in diagnosing ABPA in CF are universal. Thirteen children (10.4%) were diagnosed with sensitization to ABPA, 3 of them had aspergillosis.

Regarding the tuberculosis, only one patient (<1%) with CF had criteria for TB diagnosis.

A small percent of these children 7.25% (9 pts) were diagnosed with associated asthma.

Only one patient had active TB, bacteriological confirmed, genotype del F508 homozygous, *Pseudomonas* positive, although 16 patients (12.9%) were considered having tuberculosis, before being diagnosed with cystic fibrosis. Multiple common features present in both diseases, especially bronchiectasis , could be responsible for this.

Conclusion

ABPA is significant co morbidity in CF patients, with increasing prevalence. Although TB is a quit common condition in our country, CF children seemed to be protected against it. Further studies need to be done in order to evaluate the interrelation of cystic fibrosis with other wheezing conditions. Uniform diagnosis criteria may be useful for proper diagnosis and subsequent management.

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