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Aspergillosis in Cystic Fibrosis Patients in Russia: the results of a prospective research

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Introduction

Pulmonary aspergillosis is a serious complication in cystic fibrosis (CF) patients. According to international studies, the incidence of ABPA in CF patients ranges from 2 to 15%, and acute invasive aspergillosis is very rare. Publications on chronic pulmonary aspergillosis (CPA) in cystic fibrosis (CF) patients are limited

Objectives

The aim of the study was to evaluate the prevalence of pulmonary aspergillosis in cystic fibrosis patients.

Methods

In prospective study in 2014-2017 y.y. we included 190 CF patients. The median age was 14 y (range 1 - 37), 96 males. Total IgE test, specific *A. fumigatus* IgE and IgG tests in serum was made in all patients. Bronchoalveolar lavage (BAL) and sputum were examined with calcofluor white microscopy and culture. In case of CPA suspicion chest CT scan was performed. The diagnosis of CPA was established with D.Denning 2016 criteria, ABPA – Stevens et al. 2003 criteria, IA – EORCT/MCG 2008.

Results

Total IgE serum level varied from 1 to 3891 IU/ml (median - 149). Sensitization to fungi was identified in 57% patients. *A. fumigatus* IgG was detected in 68% patients. ABPA was diagnosed in 4,7% patients, CPA – 4,2% patients, and invasive aspergillosis – 0,5%. Severe course of CF was observed in all patients with ABPA and CPA. Patient with IA underwent liver transplantation nine months before. In ABPA patients chest CT scan abnormalities were central bronchoectases (100%), infiltrates (100%) and atelectasis (11%); in CPA patients – central bronchoectases (100%), infiltrates and focal changes (88%), and aspergilloma (11%). All patients received antifungal treatment. In ABPA patients voriconazole was used in 66%, itraconazole – 33% Median duration of antifungal treatment was 132 days (42-390), in 44% cases we observed ABPA relapse. In CPA patients voriconazole was used in 87%, posaconazole – 25% Median duration of antifungal treatment was 86 days (14-210). Overall survival in 12 months in ABPA patients was 100%, CPA patients – 87%.

Clinical symptoms in CF patients (tab. 1).

Clinical symptoms	ABPA (%)	CPA (%)
CF exacerbation	22	100
cough	100	100
hemoptysis	0	50
low lung function (FEV1% <70%)	55	63
fever	22	62
Dyspnea	100	20

Fig 4. Chest CT scan Patient A., 10 y.o., ABPA

Fig 5. Chest CT scan Patient S., 17 y.o., CPA

Fig.1. Spectrum of fungal sensitization in CF patients

Fig.2. Spectrum of aetiologic agents ABPA

Fig.3. Spectrum of aetiologic agents CPA

Conclusions

The frequency of different forms of Aspergillosis in Cystic Fibrosis patients was 9,4% (ABPA - 4,7%, CPA – 4,2%, and invasive aspergillosis – 0,5%) and requires further studies.