PS38

Alergijska bronhopulmonalna aspergiloza u bolesnika s primarnom cilijarnom diskinezijom i astmom

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KLJUČNE RIJEČI: alergijska bronhopulmonalna aspergiloza; primarna cilijarna diskinezija; astma; Aspergillus fumigatus

UVOD: Alergijska bronhopulmonalna aspergiloza (ABPA) je reakcija preosjetljivosti na spore *Aspergillus fumigatus* koja se javlja u bolesnika s astmom i cističnom fibrozom. Klinički se manifestira pogoršanjem simptoma kronične plućne bolesti uz novonastale infiltrate na plućima te razvoj bronhiektazija. ABPA u bolesnika s primarnom cilijarnom diskinezijom (PCD) je rijetka.
PRIKAZ SLUČAJA: 34-godišnji bolesnik s dijagnozom PCD i astme od djetinjstva, primljen je na bolničko liječenje zbog na CT-u toraksa vidljive tvorbe promjera 30 mm u desnom hilusu s posljedičnom atelektazom srednjeg režnja. S obzirom na radiološki nalaz, postavljena je sumnja na bronhoproliferativni proces. U laboratorijskim nalazima prati se periferna eozinofilija uz mirne tumorske markere, a u iskašljaju je izoliran *A.fumigatus* u velikom broju. Kod bolesnika je učinjena bronhoskopija, a u pristiglim citološkim materijalima pronađeni su eozinofili, hife *Aspergillusa* te Charcot-Leydigovi kristalići. Postavlja se sumnja na ABPA što se i potvrdi vrijednošću ukupnog IgE > 2000 kIU/L (do 87 kIU/L) i specifičnog IgE na *Aspergillus* 48.3 kU/L (do 0.34 kU/L). S obzirom na postavljenu dijagnozu ABPA započeta je terapija prednizolonom uz itrakonazol te kronična terapija astme i PCD. Nakon 6 tjedana terapije, prati se dobra radiološka regresija infiltrata uz opstruktivne smetnje ventilacije umjereno teškog stupnja u nalazu spirometrije i pad vrijednosti eozinofila u krvi i ukupnog IgE, međutim uz još uvijek značajne respiratorne simptome.
ZAKLJUČAK: ABPA iscrpljujuća je i kronična bolest povezana s cističnom fibrozom i astmom, međutim iznimno rijetko se nalazi u bolesnika s primarnom cilijarnom diskinezijom. Sudeći prema našem bolesniku, u ovoj skupini bolesnika liječenje ABPA je dugotrajnije i kompleksnije.

Allergic bronchopulmonary aspergillosis in a patient with primary ciliary dyskinesia and asthma

INTRODUCTION: Allergic bronchopulmonary aspergillosis (ABPA) is a hypersensitivity reaction directed against *Aspergillus fumigatus* spores that occurs in patients with asthma and cystic fibrosis. Clinically it manifests with symptoms of acute asthma exacerbation with new lung infiltrates and the development of bronchiectasis. ABPA in patients with primary ciliary dyskinesia (PCD) is rare.
REPORT: A 34-year-old patient with a diagnosis of PCD and asthma was admitted to hospital due to a chest CT scan showing a lesion 30 mm in size found in the right hilum and resulting in atelectasis of the middle lobe. Imaging findings suggested proliferative lesion in lungs. Laboratory tests detected peripheral eosinophilia along with calm tumor markers and in sputum *Aspergillus fumigatus* was isolated. Bronchoscopy was performed and in the cytological materials eosinophils, *Aspergillus* hyphae and Charcot-Leydig crystals were found. ABPA was suspected and then confirmed with the level of total IgE > 2000 kIU/L (up to 87 kIU/L) and specific to *Aspergillus* IgE 48.3 kU/L (up to 0.34 kU/L). Given the diagnosis of ABPA, prednisolone and itraconazole were prescribed along with therapy for chronic asthma and PCD. After 6 weeks of therapy radiological regression of the infiltrate was observed with moderately severe obstructive ventilatory defect and a drop in eosinophil count and total IgE levels. However, still with significant respiratory symptoms.
CONCLUSION: ABPA is a chronic disease associated with cystic fibrosis and asthma, but is very rarely found in patients with PCD. Treatment within this group of ABPA patients lasts much longer and is more complex.

KEYWORDS: allergic bronchopulmonary aspergillosis; primary ciliary dyskinesia; asthma; Aspergillus fumigatus