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PREFACE

Dear colleagues

After a long period, we have the opportunity to come together around a common interest, which is improving the treatment of patients with respiratory diseases. Actually we are in XI symposium named "Pneumology days Tešanj 2022". We are very happy that we will have the opportunity to meet our friends from the wider region, exchange experiences and learn a lot. The COVID 19 infection has entertained us all for the past two years. We were faced with great trials, we treated patients with COVID 19, but it often happened that the members of the team treating those patients got sick themselves. It was a situation where we treated both, the patients and us personally. Our motivation for learning while working was even greater. It was a challenging time in which we often witnessed treatment failure, so the learning imperative was even greater. Of course, we all tried to adequately treat the other patients as well. We hope that our theoretical and practical knowledge remained even greater in this challenge. Therefore, we sincerely look forward to these meetings and to learn a lot from each other.

Prof. Dr Besim Prnjavorac
President of Organizing Committee

ALPHA-1 ANTITRYPSIN DEFICIENCY (AATD) IN A YOUNG FEMALE PATIENT

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Introduction: The alpha-1 antitrypsin deficiency (AATD) is a hereditary autosomal codominant disease. The phenotype Pi ZZ is associated more frequently with pulmonary disease and is responsible for the presence of emphysema early in life, particularly in smokers. Generally, AATD is suspected in young patients with pulmonary emphysema or chronic obstructive pulmonary disease (COPD). Patients often suffer from diagnostic gaps and are misdiagnosed with chronic obstructive pulmonary disease (COPD), asthma, and airway hyper-reactivity (AHR), as AATD may present with nonspecific respiratory symptoms. AATD is most common in white people, and it most frequently affects the lungs and liver. In the lungs, the most common manifestation is early-onset (patients in their 30s and 40s) pan acinar emphysema most pronounced in the lung bases. However, diffuse or upper lobe emphysema can occur, as can bronchiectasis. The most frequently described symptoms include dyspnea, wheezing and cough. Pulmonary function testing shows findings consistent with COPD; however, bronchodilator responsiveness may be seen and may be labelled as asthma.

Case presentation: We describe a case of a 40-year-old Caucasian female patient, admitted to hospital because of dyspnea, malaise, cough. Symptoms started one year ago, after mild SARS-COV 2 infection. Chest X-ray during the acute illness described emphysema, with flattened diaphragm, and no signs of consolidation. According to history she was a non-smoker, office worker, with negative family history of

respiratory or liver illness. She never used any regular therapy before, no comorbid diseases and denied frequent respiratory infections during childhood. Chest computer tomography (CT) presented pan acinar emphysema most pronounced in the lung bases. Post bronchodilator spirometry revealed forced expiratory volume in 1st second (FEV₁) 54%, and forced vital capacity (FVC) 84%, with FEV₁/FVC=0.64. Routine biochemistry laboratory was normal. Gas analyses noted respiratory failure type 1 (partial) with hypoxemia partial oxygen pressure 8.1kPa, hypocapnia because of hyperventilation with partial carbon dioxide pressure 4.1kPa, and oxygen saturation 92%. Echocardiography without any findings of right heart failure, normal systolic pulmonary arterial pressure (sPAP). Abdominal ultrasound without pathological findings, no liver disease detected. According to CT finding the patient was sent to the Institute for clinical immunology and genetic disorders where the serum value of alpha-1 antitrypsin was measured. The value was 0,2 micromoles /L (reference value 5-6 micromole /L). The patient was prescribed inhaled therapy of long acting anticholinergic, short acting beta-2 agonist. She was also suggested therapy with intravenous human alpha 1-proteinase inhibitor (AAT augmentation therapy).

Conclusion: It is never too late to suspect AATD, especially in a patient with an unusual medical history. In recent years, evidence is beginning to emerge that there may be value in identifying and treating patients who do not already have deterioration of functional parameters. The Alpha-1 Foundation recommendations for the diagnosis and management of AATD in adult patients indicate that treatment should be provided for patients with FEV₁ between 30 and 65%. It may be useful to evaluate and treat patients based on clinical symptoms, even outside the established parameters, in particular cases.