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PULMONARY ALVEOLAR MICROLITHIASIS – A THIN LINE BETWEEN ORDINARY LIFE AND LUNG TRANSPLANTATION

LOVRIĆ T.[1], Hećimović A.[1], Džubur F.[1,2], Janković M.[2,1], Vukić Dugac A.[2,1], Jakopović M.[2,1], Pavliša G.[1,2], Redžepi G.[1], Samaržija M.[1,2]

[1] University Hospital Centre Zagreb, Zagreb, Croatia
Department for Respiratory Diseases Jordanovac

[2] University of Zagreb, Zagreb, Croatia
School of Medicine

Introduction: Pulmonary alveolar microlithiasis (PAM) is a rare, diffuse lung disease characterized by deposition of calcium phosphate within alveolar airspaces. Since the first description of the disease 150 years ago, more than 1000 cases have been reported in the world literature. PAM is often discovered on chest radiographs obtained for other reasons during early adulthood (from birth up to 40 years of age). Patients typically remain asymptomatic until middle age, when pulmonary fibrosis, pulmonary hypertension, and chronic respiratory failure ensue. Chest radiographs reveal diffuse, hyperdense, micronodular shadows producing a characteristic snowstorm appearance. The recent discovery in patients with PAM of genetic mutations in the SLC34A2 gene, which encodes the sodium-phosphate cotransporter NPT2b, has opened a window into PAM disease pathogenesis.
Case report: A 28-year-old female with a history of PAM was transferred to our clinic on the 27th January 2016 on mechanical ventilation due to sepsis and bilateral pneumonia. The patient had been diagnosed at the age of 9 and was asymptomatic until the age of 28. She conceived and gave birth in November 2015. She presented on 8th January 2016 with fever, cough without sputum, fatigue and shortness of breath. At first she was hospitalized at General Hospital but was transferred to Mostar University Hospital due to worsening of her respiratory insufficiency. On the 17th January she was intubated and the mechanical ventilation was initiated.
At the time of admission at our Clinic she had elevated inflammatory parameters, radiological characteristics of

PAM with bilateral pneumonia. She was treated with broad-spectrum antibiotics, which were modified due to microbiological results of bronchoalveolar lavage (Staphylococcus aureus MRSA, Pseudomonas aeruginosa and Acinetobacter baumannii). We started urgent lung transplantation evaluation process. Fortunately, she responded well to treatment with the decrease in inflammatory parameters and improvement of respiratory insufficiency. On the 11th February she was tracheotomised and a few days later extubated and ever since breathes independently. From the June 2016 she is on the active lung transplantation waiting list. Her last reevaluation at our Clinic was in April 2017 with stable pulmonary function and good exercise tolerance. Any respiratory infection may lead to worsening of her respiratory insufficiency and fatal outcome.
Conclusion: There is no known medical treatment to reduce or halt the progression of PAM. The only effective treatment remains lung transplantation.
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