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Diffuse Cystic Lung Disease Diagnosed in Adolescence Period

Tuğba Ramaslı Gürsoy¹, Zeynep Reyhan Onay¹, Tuğba Şişmanlar Eyüboğlu², Ayşe Tana Aslan¹, Öznur Boyunağa³, Leyla Memiş⁴, İrfan Taştepe⁵

¹Department of Pediatric Pulmonology, Gazi University School of Medicine, Ankara, Turkey

²Clinic of Pediatric Pulmonology, Dr Sami Ulus Maternity and Children Training and Research Hospital, Ankara, Turkey

³Department of Pediatric Radiology, Gazi University School of Medicine, Ankara, Turkey

⁴Department of Pathology, Gazi University School of Medicine, Ankara, Turkey

⁵Department of Thoracic Surgery, Gazi University School of Medicine, Ankara, Turkey

Introduction: Diffuse cystic lung disease (DCLD) is a heterogeneous group of diseases characterized by air-filled, thin-walled, rounded or irregular-edged cavities in the lung parenchyma. Pneumothorax is the most common manifestation of DCLD and may also appear as a symptom of presentation. In this report, we present a case of DCLD diagnosed in adolescence with spontaneous pneumothorax.

Case Presentation: A 17-year-old male patient was admitted to pediatric pulmonology department with the complaint of chest pain, shortness of breath. The patient had a history of twice hospitalization, chest tube replacement due to pneumothorax in last month and no previous lung disease, cigarette smoke exposure and family history of pneumothorax. His weight and height were 3-10 percentile. His oxygen saturation was normal. Increased anteroposterior chest diameter and decreased breathing sounds in bilateral lower lobes were found in physical examination. The laboratory tests were normal except increased C-reactive protein (73 mg/dL). Antinuclear antibody detected as weak positive, fine speckled pattern. In thorax tomography, air cysts were common in both hemithorax and minimal pneumothorax in the right lung. His dermatological and ocular examination, radiographic skeletal survey, abdominal ultrasonography and cranial MR were normal. Unilateral pleurodesis and lung biopsy were performed. The pathology of the patient's lung biopsy material was reported as pulmonary parenchyma characterized by widespread emphysematous changes, focal atelectasis and congestion. Diseases other than Birt-Hogg-Dubé syndrome (lymphangioleiomyomatosis, pulmonary Langerhans cell histiocytosis, lymphocytic interstitial pneumonia, follicular bronchiolitis, and light-chain deposition disease) were excluded by these findings. FLCN gene mutation result is expecting.

Conclusion: DCLD is a rare disease in childhood. Although under the age of 18 years, as a childhood age group, adolescent patients should be evaluated in terms of the causes of adult DCLD. Clinicians should review the differential diagnoses in a multidisciplinary manner for the diagnosis of DCLD.

Keywords: Adolescence, diffuse cystic lung disease, pneumothorax