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A Rare Diagnosis in Childhood: Idiopathic Giant Bullous Emphysema

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Introduction: Idiopathic giant bullous emphysema (Vanishing lung syndrome) is a disease with bullae covering at least 1/3 one or both hemithorax. It is a disease characterized by dyspnea and spontaneous pneumothorax in younger and smoker male patients. We aimed to present a case with the diagnosis of VLS because of the small number of children reported in the literature.

Case Presentation: A 15-year-old girl, who underwent peritoneal dialysis with nephrectomy because of acute graft rejection following renal transplantation due to tubulointerstitial nephritis two years ago, was referred because of decreased breathing sounds in the right hemithorax on physical examination. The patient had no complaints. She was not smoking but she was exposed to cigarette smoke. On examination she was not hypoxic but her respiratory sounds were decreased in the right hemithorax. Chest radiography was interpreted as showing a hyperlucent right hemithorax with a left shift of the mediastinum. Spirometry revealed severe restrictive pattern. Chest computerized tomography showed widespread emphysematous changes of bullous features related to all of the right lung, pushing to the left in mediastinal structures, and focal emphysema areas in the left posterobasal segment. Further investigation results included a normal α_1 -antitrypsin concentration, normal sweat chloride titration and negative autoantibodies (ANA and ANCA). Bronchoscopy revealed bronchial lavage smears with non-common mucinous metaplasia. Therefore lung biopsy was performed in the lower lobe of the right lung. Histopathological examination revealed bullous changes of lung and K-Ras mutation was not found. She was diagnosed with "Idiopathic giant bullous emphysema". No surgical intervention could not be performed because the patient did not continue to follow up.

Conclusion: Although idiopathic giant bullous emphysema is a rare disease in childhood, it should be kept in mind that bullous emphysema is detected in children.

Keywords: Bullous, childhood, emphysema