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Immunodeficiencies in childhood interstitial lung diseases: First data from the Turkish National chILD registry

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Abstract

Objective: Immunodeficiencies are associated with different Childhood Interstitial Lung Diseases (chILD) patterns, but data in this area are limited. We aimed to reveal the diagnoses, lung involvement, and clinical characteristics of immunodeficiency patients diagnosed with chILD.

Method: All patients diagnosed with immunodeficiency(Diagnosis code:B3) registered in the Turkish Registry of chILD in 2023 were included. Age at presentation, age at diagnosis, type of immunodeficiency, clinical, laboratory, and radiological findings, and clinical status of patients were noted.

Results: A total of 667 patients were recorded into the registry, and 114 (17%) had immunodeficiency. The mean age of patients with immunodeficiency was 12,4±6.1 years and 47% of them were female. Among the patients, 60% had primary immunodeficiency; the others were secondary, mainly secondary to hematopoietic stem cell transplantation (HSCT). The most common radiological finding was ground glass opacity, nodules, bronchial thickness, and linear opacities, respectively. The most common diagnosis was bronchiolitis obliterans after HSCT and chILD without any specific diagnosis. However, there are many different diagnoses, such as GLILD, PAP, pleuropulmonary fibroelastosis, hypersensitive pneumonitis, etc. The mean FEV1 % was 58.5±28.7,and FVC 61.3±27%. Seven of them died during follow-up.

Discussion: Immunodeficiencies are an important cause of chILD. Patients may present with different types of involvement. Mortality and morbidity may be common in children with immunodeficiency and chILD. As registries become more widespread, knowledge on immunodeficiency and chILD will also increase

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Abstract

Introduction: Despite the great impact of PIBO on children, clinical data is scarce, and most clinicians need more awareness of the disease. We aimed to evaluate patients' demographic, clinical, radiologic findings with PIBO in the Childhood Interstitial Lung Diseases Registry of Türkiye(chILD-TR).

Materials and Methods: The current age, gender, symptoms, systemic diseases, physical examination, laboratory and radiological findings, therapies of patients with PIBO in the chILD-TR database were recorded in 2022-2023.

Results: There were 180 patients, 61.1% were male. The current age of the patients was 103.23(13-364), the age at which symptoms started was 25.96(0-233.1), the age at hospital admission was 48.65(0-303.9) months.56.1% of the patients had a history of recurrent lower respiratory tract infection, 17.8% of the patients had growth retardation. The most common symptom was cough(77.8%). Oxygen was used in 12.8% of the patients, noninvasive mechanical ventilator was used in 3.3%. The most common physical examination finding was rhonchi(61.7%). There was a mosaic perfusion pattern in all patients on thorax CT;other common findings were increased aeration(42.8%), thickening of the bronchial wall(41.1%). Mean FEV1, FVC, MEF25-75 values were 61.12±21.68%, 66.45±22.55%, 50.26±28.10%. The most common infection agent was adenovirus(17.2%). 32.2% of patients were using oral steroids. The follow-up period was 85.24±68.29 months. 2 of the patients died.

Discussion: Physicians need better awareness of PIBO as patients were referred to a pediatric pulmonologist almost 2 years after symptom onset. Patient registries have an essential role in increasing awareness.

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