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Türkiye'den gerçek yaşam verilerine göre, bağışıklığa ilişkin sorunlar önemli klinik sorunlara yol açmamıştır.

Kilavuz S et al. Real-world patient data on immunity and COVID-19 status of patients with MPS, Gaucher, and Pompe diseases from Turkey. Arch Pediatr 2022;29:415-23.

Background: COVID-19 and lysosomal storage disorders (LSDs) share a common immunological pathway as they cause the release of cytokines in a similar pattern. We aimed to evaluate the immunity status and reveal the course of COVID-19 in patients with LSDs.

Results: The median age of 110 patients with LSDs was 129 months (range: 21-655), and all but one patient with mucopolysaccharidosis (MPS) type III were regularly receiving enzyme replacement therapy (ERT). In 53.6% (n = 56) of the MPS [8 type VI, 11 type IVA, 1 type III, 3 type II], and 7 patients with Gaucher disease, an abnormality in at least one of the autoimmunity or immunodeficiency parameters was reported. Furthermore, 12 (57%) of 21 Gaucher cases (7 type III, 5 type I), 18 (40.9%) of 44 MPS cases (9 type IVA, 5 type VI, 1 type I, 2 type II, and 1 type III), and six (66%) of nine Pompe cases were reported to involve abnormalities in at least one of the parameters related to immunodeficiency. Immunoglobulin (Ig) M and IgA levels were reported to be lower, and there were abnormalities in the lymphocyte counts and subgroups in the MPS group. ANA was reported to be positive in one patient with Gaucher type III, anti-DNA in two patients with Gaucher type I and one patient with MPS type VI, antithyroglobulin in two patients with Gaucher type I, anti-TPO in one patient with Gaucher type I, TRAB in one patient with Gaucher type I, antiphospholipid IgM in three patients with Gaucher type I, and one patient with Gaucher type I, anticardiolipin IgM in one patient with Gaucher type I, one patient with Gaucher type III, and one patient with MPS type II. However, no clinical presentation was consistent with the laboratory results except for one patient with Gaucher type I disease with Hashimoto thyroiditis. Two of the four patients who survived the COVID-19 infection with mild symptoms had a diagnosis of Gaucher type I, and no abnormality was detected in their laboratory tests. The other two patients had a diagnosis of MPS types VI and II. Immune dysfunction was detected in the patient with a diagnosis of MPS type II. Four of our patients were discharged without any sequelae.

Conclusion: Problems with immunity did not cause any noticeable clinical results. Being well protected by reducing social contact might have played a role. However, we believe that it should be borne in mind that cardiac and pulmonary involvement, as well as immune dysfunction in LSDs, may cause an increased need for intensive care because of secondary bacterial infections.

