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SCIENTIFIC LETTER

Pneumocystis jirovecii and SARS-CoV-2 Coinfection as Presentation of X-linked Severe Combined Immunodeficiency

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To the Editor: A 6-mo-old boy was admitted to the Pediatric Intensive Care Unit due to severe hypoxemia secondary to bilateral pneumonia, with a positive SARS-CoV-2 PCR in the nasopharyngeal swab. He developed a pediatric acute respiratory distress syndrome (PARDS) requiring endotracheal intubation within the first hours after admission. The transthoracic echocardiography showed no signs of pulmonary hypertension or congenital heart disease. Due to the atypical clinical course and recent history of persistent cough and weight loss, a complete immune function test was performed, showing absence of CD3+ cells and CD3-CD16/CD56+ cells with normal CD19+ cell count, but associating hypogammaglobulinemia and absence of memory B cells, compatible with a T−B+NK− severe combined immunodeficiency (SCID) phenotype. Given the possibility of a *Pneumocystis jirovecii* pneumonia (PJP), empirical treatment with intravenous trimethoprim-sulfamethoxazole was started, with a subsequent positive result for *P. jirovecii* in a tracheal aspirate sample. He also received methylprednisolone (2 mg/kg/d). The patient's status progressively improved, receiving 4 wk of trimethoprim-sulfamethoxazole treatment and being extubated after 5 wk. The next-generation sequence study of the 200 immunodeficiency gene panel identified a frameshift variant in hemizygosis (p. Ser113LeufsTer34) in exon 3 of the *IL2RG* gene, compatible with the clinical manifestations and phenotype of the patient. He was finally transplanted from a matched unrelated donor four months after the diagnosis of SCID, with a good outcome.

Severe Immunodeficiency and Coinfection must be Ruled out in Pediatric Patients with Severe SARS-CoV-2 Infection [1]. Patients with X-linked SCID may present with opportunistic infections, especially PJP [2]. Both PJP and SARS-CoV-2 infection can trigger PARDS, defined by a respiratory failure with pulmonary edema not explained by cardiac failure or fluid overload [3]. Trimethoprim-sulfamethoxazole should be empirically started as soon as SCID is suspected, before microbiological confirmation, as it might improve the outcome [4]. Corticosteroids are also essential for PJP treatment [2].

Declarations

Conflict of Interest None.

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