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Letter to the Editor

Haemolytic anemia triggered by SARS-CoV-2 in patient with hereditary spherocytosis[☆]



Anemia hemolítica desencadenada por SARS-CoV-2 en una paciente con esferocitosis hereditaria

Dear Editor,

Hereditary spherocytosis (HS) is a congenital haemolytic anaemia characterised by an alteration in the erythrocyte membrane, mainly due to defects in the membrane proteins ankyrin, band 3 and spectrin. This leads to a loss of cohesion of the lipid layer of the red blood cell and a decrease in its surface area, resulting in the formation of the disease's characteristic spherocytes, which show reduced deformability and are consequently destroyed as they pass through the spleen.¹

It is a disease with great clinical heterogeneity, with asymptomatic cases (compensated haemolysis) and others with haemolytic anaemia of varying degree requiring regular transfusions. On occasions, and especially after viral infectious processes, haemolytic crises occur that may require transfusion of packed red blood cells or in which their requirement increases.¹

Treatment of HS patients is supportive; splenectomy is usually reserved for the most severe cases.²

We report the case of a 4-year-old girl, diagnosed with HS at 15 months of age, with a family history of the same disease (her mother). The patient had mild HS, without anaemia, jaundice or splenomegaly and no haemolytic crises during infectious processes.

The patient came to the emergency department with a 48 h-history of 39 °C fever, together with excess mucus, food refusal and significant mucocutaneous jaundice. In addition to jaundice, the physical examination revealed a 3-finger breadths splenomegaly. Cardiopulmonary auscultation and the rest of the examination were normal. The family did not report known contacts with patients affected by COVID-19.

A complete lab test was requested, which showed a haemoglobin of 9.3 g/dL, reticulocytes of $0.51 \times 10^6/\text{mm}^3$; bilirubin of 5.93 mg/dL; LDH of 1835 IU/L and negative direct Coombs. A SARS-CoV-2 PCR was requested and found to be positive. A contact study was carried out, in which all cohabitants tested positive, and it was decided to admit the patient for close monitoring in case of haemolytic crisis.

In subsequent days, progressive anaemia occurred until a haemoglobin of 7.4 g/dL was reached, accompanied by parameters of haemolysis and splenomegaly, together with persistence of fever up to 39 °C during the first 3 days of admission, despite the decrease in acute phase reactants. The patient was hemodynamically

ically stable, improving the initial excess mucus and no signs of respiratory distress, so a watchful waiting approach was decided, without transfusing packed red blood cells or initiating antibiotic therapy.

On the 5th day of admission, an improvement in haemoglobin levels was detected, together with a decrease in haemolysis parameters, a decrease in jaundice and clinical improvement, so it was decided to discharge the patient and monitor her on an outpatient basis.

One week later the patient's haemoglobin levels were within normal range, with signs of mild haemolysis and resolution of the splenomegaly. To date, the patient remains asymptomatic, without having developed new complications.

In patients with HS, anaemia episodes can be observed for several reasons: haemolytic crisis after viral infections, aplastic crisis due to parvovirus B19 or megaloblastic crisis due to folic acid deficiency.¹

Haemolytic crises are one of the most common complications, generally in the context of feverish symptoms due to viral infection of any type, although they can appear spontaneously. They present with a worsening of the usual anaemia, jaundice, increased markers of haemolysis (reticulocytes, LDH . . .) and splenomegaly, as our patient. In these cases, close monitoring of patients is especially important, since the symptoms and intensity of haemolysis can range from mild to moderate-severe symptoms.^{1,3}

The case reported highlights the importance of closely monitoring patients at risk of haemolytic crisis during the pandemic due to the SARS-CoV-2 virus infection. To date, several studies have been published on haemolytic crises in patients with sickle cell disease and those affected by COVID-19⁴; however, there is little mention of the risk associated with patients with membrane haemoglobinopathies infected by this virus.

In the reviewed literature (PubMed search term: *hereditary spherocytosis AND COVID-19*, in the years 2020–2021) we have only found 2 reports of cases and only one of them in paediatric age.^{3,5}

In the context of the SARS-CoV-2 pandemic, it would be important to detect and monitor, in a concise manner, signs of haemolysis in patients with haemolytic anaemias due to membrane defects affected by COVID-19, even if they initially present with mild symptoms. In addition, as in the case of our patient, haemolysis in patients with HS and COVID-19 can be of great intensity, even if they present mild underlying symptoms or are even asymptomatic.

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