Cow’s milk protein intolerance imitating septic shock in a young infant

Intolerancia a la proteína de la leche de vaca simulando shock séptico en un lactante pequeño

Dear Editor,

Food protein-induced enterocolitis syndrome (FPIES) is the most serious form of enteropathy. The pathophysiology of this syndrome is not completely understood, but it is believed that ingestion of the triggering food is followed by T-lymphocyte activation, resulting in a local inflammatory response, increased gastrointestinal (GI) permeability and subsequent passage of fluids to the GI lumen.1

A boy aged 4 months visited the emergency department (ED) with profuse vomiting and signs of hypovolaemic shock. He had been breastfed until the age of three months, when supplementation with one feed of cow’s milk formula a day was introduced. In the two days that followed, he vomited once a day right after ingesting the cow’s milk. On day 3, he developed persistent vomiting and bloody diarrhoea and became increasingly lethargic.

In the ED, he presented in shock, with a blood pressure of 60/25 mmHg, poor peripheral perfusion with cold extremities, and no other abnormalities in the physical examination.

The diagnoses considered at this point were sepsis and cow’s milk protein allergy.

References

The blood tests showed absence of leucocytosis (10,990/μL) with relative neutrophilia (76.2% neutrophils), thrombocytosis (645,000/μL platelets), a negative result for C-reactive protein, normal levels of serum electrolytes and normal renal function. There were no abnormalities in the cerebrospinal fluid or the urine.

The boy was given a bolus of isotonic fluid (20 mL/kg) to reverse the shock, intramuscular adrenaline and methylprednisolone. Antibiotic therapy was initiated with ceftriaxone.

The patient had normal total IgE levels, and negative results for specific IgE against cow’s milk protein. The trip-tase value measured at the emergency department was also normal.

Once admitted to the paediatrics ward, he was fed with breast milk or with an amino acid-based formula, with complete resolution of symptoms in less than 12 h. All the culture results were negative.

The compatible history and favourable clinical outcome allowed the conclusion that the patient had cow’s milk protein-induced enterocolitis syndrome (MFPIES).

Two months after the acute event, the patient underwent an oral food challenge (OFC) with extensively hydrolysed milk formula (EHF), with recurrence of symptoms, but at the age of 12 months he tolerated this formula during a new OFC.

At the age of 2, under medical supervision, the patient underwent oral tolerance induction. The starting dose was 1 mL, which was doubled weekly until reaching a 200 mL dose. The patient exhibited good tolerance.

The clinical manifestations of FPIES and its severity are dose-dependent. In this case, the identification of cow’s milk proteins as the trigger was confirmed by the previous history of exposure to small amounts followed by reactions of increasing severity. The diagnosis of acute FPIES requires that a patient meets the major criterion:

- Vomiting in the 1–4 h period after ingestion of the suspect food in the absence of classic IgE-mediated allergic skin or respiratory symptoms;

and at least 3 minor criteria:

- A second (or more) episode of repetitive vomiting after eating the same suspect food
- Episode of repetitive vomiting 1–4 h after eating a different food
- Extreme lethargy with any suspected reaction
- Marked pallor with any suspected reaction
- Need for emergency room visit with any suspected reaction
- Need for intravenous fluid support with any suspected reaction
- Diarrhoea within 24 h
- Hypotension
- Hypothermia

This infant fulfilled the major criterion and all of the minor criteria with the exception of hypothermia. He also had thrombocytosis, which has been reported in 65% of patients with acute FPIES, and relative neutrophilia, a common laboratory finding in patients with positive oral food challenges (OFC). In patients with a single documented episode, performance of an OFC can be considered. In this case, we considered that the typical history made it unnecessary.

As occurs in IgE-mediated cow’s milk allergies, where reactions may be triggered by an EHF, about 10–40% of patients with MFPIES do not tolerate these formulas and require formulas based on amino acids, although their financial and nutritional impact must be taken into account.

There are geographic and institutional variations in the timing of OFC in patients with FPIES. Most authors defend that it should not be tried before 12–24 months have elapsed since the last acute episode, but good results have been observed after 10 months in patients with MFPIES. It is important to emphasize that regardless of the timing, OFCs should always be performed in appropriate settings, where resuscitation equipment and staff trained in the identification and treatment of these potentially severe reactions are always available.

In conclusion, knowledge of this syndrome and the severity of its presentation allows the early removal of the trigger and prevents unnecessary testing and pharmacological treatment. The follow-up and management of these patients can be challenging, but the disorder has a favourable prognosis and resolves fully in most cases by age of 3–5 years.

References


Sylvia Jacob a, Artur Bonito Vitor b
a Departamento de Pediatría, Centro Hospitalar Sáo João, Oporto, Portugal
b Departamento de Inmunología Pediátrica, Centro Hospitalar Sáo João, Oporto, Portugal

* Corresponding author.
E-mail address: sylviamhelder@gmail.com (S. Jacob).

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