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Case Report

A Case Report of Acute Food Protein-Induced Enterocolitis Syndrome that Challenges the Existing Diagnostic Consensus

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Abstract

Acute food protein-induced enterocolitis syndrome (FPIES) is a food-related gastrointestinal hypersensitivity disorder. It is classified as a non-IgE-mediated syndrome, but some atypical forms may have a slight IgE increase. It is characterized by repetitive vomiting within 1 - 4h after ingestion of a trigger food, in association with pallor, lethargy, diarrhea, hypotension or hypothermia. The current consensus guidelines of acute FPIES diagnosis requires an absence of classic IgE-mediated allergic skin or respiratory symptoms in this syndrome, but more evidence has surged that may contradict this initial definition. We present an unusual case of acute FPIES in a six-month-old infant, that besides the classic display, also includes a skin manifestation and a slight IgE positivity to the trigger food, potato. This case report highlights the need to review criteria for FPIES diagnosis.

Keywords: *Diagnosis; Food Allergy; Food Protein Induced Enterocolitis Syndrome; Potato; Skin Manifestations*

Abbreviations

FPIES: Food Protein-Induced Enterocolitis Syndrome; OFC: Oral Food Challenge

Introduction

Food protein-induced enterocolitis syndrome (FPIES) is a food-related gastrointestinal hypersensitivity disorder [1-4]. It is believed to be a non-IgE-mediated allergy, but atypical cases may present a slight specific IgE increase [1-4]. Its pathophysiology is still poorly understood, but it seems that food proteins induce a local inflammation that increase intestinal permeability with fluid passage into the intestinal lumen [1]. Since it is a little-known syndrome, with little evidence to date, an International Consensus Guidelines for the diagnosis and management of FPIES was published in 2017 [1]. These guidelines describe acute FPIES as repetitive vomiting episodes, presented within 1 - 4h from the triggering food ingestion without classic IgE-mediated allergic skin or respiratory symptoms (major criterion), that may

be accompanied with minor criteria (≥ 3) such as marked pallor, extreme lethargy, diarrhea (< 24h) and, in more severe cases, dehydration with hypotension, hypothermia and need for intravenous fluid support [1]. This reaction usually resolves over a matter of hours [1]. Without the offending food, the patient is usually asymptomatic showing a normal growth and development [1].

In this consensus, the presence of classic IgE-mediated allergic skin symptoms is an excluding factor of acute FPIES diagnosis [1], but the authors aim to report a case of acute FPIES with skin manifestations.

Case Report

A six-month-old girl, with an irrelevant medical history, was brought to our hospital for skin lesions after the introduction of solid foods.

She was breastfed for the first four months of life, in combination with milk formula from two months, passed on to complementary feeding at four and a half months of age, when she started to ingest soup (potato, carrot, and onion). Four hours after the ingestion of the first one, perioral and neck rash was observed. The following day the same symptoms were observed with the same soup. Complementary feeding was interrupted for a few days without any symptoms and reintroduced again with soup (potato, carrot, onion, and pumpkin), with the same rash occurring four hours later. Complementary feeding was suspended once again for a week, without any reactions during this period. Later, soup (potato, carrot, onion, and chicken) was reintroduced again. This time she developed a rash after three hours, and besides perioral and neck involvement, she also presented with hand and chest rash. A few days later, with another asymptomatic break in between, she was given another soup (potato, carrot, and chicken) with the development of the same perioral and neck rash three hours later (Figure 1).

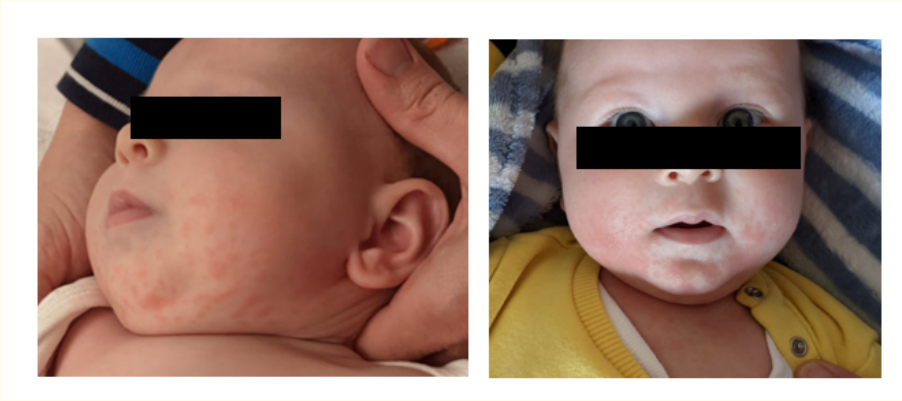

Figure 1: *Skin symptoms in two different episodes of food allergy.*

The diagnostic hypothesis of an oral allergic syndrome was raised, however due to its unusual late onset, we decided to perform an oral food challenge (OFC) with soup (potato, carrot, onion, and pumpkin). After three hours the patient showed neck and chin rash. But surprisingly, after four hours she also began profuse vomiting (> 10 episodes) associated with nausea, hypotonia and extreme lethargy, pallor, sleepiness, and late diarrhea.

Although the clinic was very suggestive of acute FPIES, as we also could be facing an anaphylaxis (cutaneous and gastrointestinal involvement), adrenaline (0.1 mL 1:1000, intramuscular via) was administered, yielding no effect on skin lesions, hypotonia, lethargy, pallor, or drowsiness. The infant was hospitalized with ondansetron and fluid therapy, with clinical discharge in the next day, asymptomatic.

Therefore, a more detailed investigation was subsequently carried out. A skin prick test and a prick-prick were performed that showed positive for histamine (8 x 5) and raw potato (4 x 3) and negative for boiled potato, potato extract, raw and boiled carrot, carrot extract, raw and boiled onion, onion extract and raw and boiled pumpkin. She had a total IgE of 9 Ul/ml and specific IgE for potato of 1.16 kU/L.

Afterwards she maintained a potato-free diet, though accidental contact occurred at eight months of age, resulting in another episode of acute classic FPIES with multiple vomiting, prostration, sleepiness, and lethargy, but this time without skin symptoms.

At two-years-old an OFC with potato was performed, which was positive, causing an episode of acute classic FPIES, but less severe than the first one.

Currently potato eviction is maintained without further episodes. She ingests other vegetables with tolerance, including other Solanaceae such as tomato and eggplant.

Discussion and Conclusion

This case described an infant presenting with a rash after every meal, at the beginning of complementary feeding. She was asymptomatic on every interruption. This initial manifestation did not point to an acute FPIES, since the five times she ate soup at home, she only developed a skin rash, without further symptoms.

However, with the OFC with soup, the infant showed symptoms suggestive of acute FPIES, fulfilling six minor criteria (multiple episodes with the same food, extreme lethargy, marked pallor, diarrhea, need for assistance in an emergency department and intravenous fluid support) and partially the major criterion (she had 10 vomits 4h after ingestion, but she did not have the required absence of skin symptoms). Despite the gastrointestinal and skin involvement, this case was different from an IgE-mediated anaphylaxis which usually occurs within a few minutes or maximum one to two hours, after the ingestion of the offending food [3].

Even though we consider an acute atypical FPIES the most appropriate diagnosis for this case, since the infant was slightly sensitized to potato, we also believe that it is a very peculiar case of acute FPIES because of the coexistence of skin symptoms (at the moment of the OFC).

It is not the first time that skin manifestations have been described in acute FPIES cases [2,3]. According to the experts of the International Consensus Guidelines, published in 2017, allergic IgE-mediated skin symptoms would exclude acute FPIES diagnosis in this infant. But they also assess as a future need, the priority to validate the proposed diagnostic criteria. For this reason, and in accordance with some other articles [2-5], we believe that this part of the major criterion could be reviewed in the next update.

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Conflict of Interest

The authors have no conflicts of interest relevant to this article to disclose.

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