

CASE REPORT

Egg provoked food protein-induced enterocolitis-like syndrome in an adult

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Key Clinical Message

Food protein-induced enterocolitis syndrome is a non-IgE-mediated food allergy usually diagnosed in infancy. We report a case of a similar syndrome in an adult, following ingestion of egg. We remind clinicians to consider this diagnosis which may present to emergency physicians and gastroenterologists long before an allergist is consulted.

Keywords

Egg allergy, enterocolitis, food allergy, food protein-induced enterocolitis syndrome, non-IgE-mediated food allergy.

Food protein-induced enterocolitis syndrome (FPIES) is an uncommon but serious disorder and has been defined as affecting infants with onset in the first year of life [1]. Cow's milk and soy are the most frequent causative foods, and the natural history is usually one of spontaneous resolution in childhood [2]. We report a case consistent with FPIES of adult onset and attributable to egg ingestion.

A 43-year-old female care assistant presented for assessment of a two-year history of recurrent episodes of vomiting and diarrhea with abdominal pain. Each episode onset occurred several hours after eating eggs, and on four occasions, her symptoms were severe enough to warrant presentation to an emergency department (ED). Clinical symptoms were restricted to the gastrointestinal tract. On one admission, renal impairment was evident, with an elevation of the serum creatinine to 83 (normal range 44–80 mmol/L), with subsequent normalization to 56 mmol/L, suggesting significant dehydration. Hypotension was not documented. A peripheral blood neutrophilia of $12.6 \times 10^9/L$ (normal range 2.0–7.5) was simultaneously present, which later normalized to $5.9 \times 10^9/L$. Fecal cultures and toxin detection were negative, but a stool leukocytosis was evident on microscopy. On these occasions, she was treated with intravenous fluid

resuscitation with benefit. Several milder episodes had occurred after eating omelet, meringue, and salad containing boiled eggs with diarrhea and abdominal pain not requiring ED attendance. Once the patient had identified this pattern herself, she strictly followed an egg-exclusion diet for several months with absence of all gastrointestinal symptoms.

Skin prick testing revealed negative responses to egg white and yolk, and serum-specific immunoglobulin E (IgE) to both egg white and yolk were similarly negative. No alternative causes for her episodes became apparent following gastrointestinal review, including abdominal CT scan and endoscopies with macroscopically normal colonic mucosa at a time of dietary egg avoidance. A clinical diagnosis of adult-onset FPIES attributable to egg was made. Eighteen months after successful egg avoidance, our patient attended a family party and ate a piece of cake (containing baked egg), waking at 4.30 AM the next morning with diarrhea and colicky abdominal pain. On the basis of this event, the patient did not consent to perform a supervised challenge. The patient was advised to continue her previously successful avoidance of all egg-containing foods. An adrenaline auto-injector was not prescribed.

Food protein-induced enterocolitis syndrome -like syndromes in adults are not well defined although there has

been a case report implicating scallop [3] and a case series in eight adults related to various shellfish [4]. Tan *et al.* reported 31 adults with a variety of gastrointestinal symptoms that had occurred after food on at least two occasions, with a variety of foods mostly seafood and egg; some of these may represent FPIES-like syndromes [5]. Caubet reported 160 cases of FPIES, predominantly in children, with only 13 of the patients having a diagnosis after the age of 5 years, and at least a few of which were adults [6]. Ruffner *et al.* included two adolescents in their large series of 462 patients, who retained the reactivity from infancy [7]. De novo onset in adults, therefore, appears uncommon, although there may be individuals reactive to shellfish, for example, who simply avoid the food and never present for assessment.

It remains to be seen whether a similar syndrome in adults has comparable pathophysiology to that seen in infants, although even this is incompletely understood [8] and is likely to be T-cell dependent [9]. The NIAID criteria for diagnosis do not require histological evidence. However, clarification of the acute pathology in adults will be a critical feature to better define the diagnosis and progress understanding of any pathophysiologic mechanism. Importantly, the clinical course of infants is usually one of spontaneous resolution; the timing may vary according to the culprit food [10]. Other than as a diagnostic procedure, the second role of oral challenge testing is to establish resolution of the condition, highly relevant in children who may then resume a normal diet [11]. It is unknown whether remission might naturally occur in adults, but is worthy of consideration. Guidelines for diagnosis of the condition emphasize clinical features, mainly relating to infancy, and the gold standard of an oral food challenge [12]. Repeated presentations to the ED are typical, and in Ludman's case series, most patients presented more than once before the diagnosis was suspected or made [13].

Egg is far from the most common food associated with FPIES in published series [1, 14], although Ruffner *et al.* [7] found egg to be relevant in 11% of their 462 patients and Ludman *et al.* [13] in 13% of their 54. Whether or not lightly cooked egg versus well-baked egg-containing foods makes a difference, as it can do in IgE-mediated egg allergy, is not known. In our patient, symptoms were elicited even after ingestion of cake baked with egg.

The patient had previously received the influenza vaccine in the course of her employment within a hospital. She had deferred the vaccination in recent years due to concerns about egg content. Given that this remained indicated for her in her workplace, she was administered a dose of Vaxigrip influenza vaccine (Sanofi Pasteur SA, 2015 ovalbumin content <0.05 mcg/0.5 mL dose) under our supervision without adverse effect.

Food protein-induced enterocolitis syndrome is a well-defined entity in infants, with a typical natural history of spontaneous resolution. Diagnosis is confirmed with a supervised oral challenge, although NIAID guidelines for FPIES state that if there has been "hypotension or multiple reactions to the same food" that resolution of symptoms with an elimination diet is sufficient [12]. The syndrome is not often diagnosed in adults and the long-term prognosis is unknown. We report this clinical case, as we believe it is most consistent with an adult-onset FPIES-like syndrome. Egg is an uncommon causative food in infant FPIES series. Administration of the influenza vaccine, now known to be safe in those with IgE-mediated egg allergy, including anaphylaxis, was tolerated in our patient. Awareness of the possibility of this syndrome in adults, including the difficulty of establishing a firm diagnosis, calls for careful clinical assessment. Identification of further cases may facilitate better definition and diagnostics for this potentially severe syndrome.

Conflict of Interest

None declared.

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