



Clinical Notes

Progress of telangiectasia macularis eruptiva perstans exacerbated egg allergy

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Telangiectasia macularis eruptiva perstans (TMEP), a form of cutaneous mastocytosis, rarely occurs in childhood and is associated with systemic involvement in 45% of adult cases.¹ Multiorgan involvement causes systemic symptoms, including pruritus, flushing, or abdominal pain due to mast cell degranulation and subsequent release of chemical mediators.² We report a pediatric case of TMEP presenting with immunoglobulin E (IgE)-mediated egg allergy, which worsened as the skin lesions proliferated.

A 10-year-old girl was referred to our hospital for oral immunotherapy initiation. She was asthmatic but had no history of eczema. Both parents had been allergic to hens' eggs as children, and the mother had atopic dermatitis. At 1 year of age, redness of the skin was observed upon contact of egg soup with the patient's face. She was diagnosed with hens' egg allergy following testing with 20.80 kUa/L of egg-white-specific IgE (ImmunoCAP, Thermo Fisher Scientific Japan, Tokyo) and completely avoided egg products thereafter. At 2 years of age, she accidentally ate dumplings containing egg whites, which resulted in vomiting. On oral food challenge test (OFC) at 5 years of age, after ingesting 0.5 g of a whole boiled egg, she developed anaphylaxis manifesting as multiple episodes of vomiting and wheezing, which required adrenaline administration. At 6 years of age, serum-specific IgE analysis showed the following: egg yolk, 0.65 kUa/L; egg white, 4.89 kUa/L; and ovomucoid, 3.46 kUa/L. On OFC, she was able to consume one boiled-egg yolk. Throat itching was observed after consumption of 0.5 g egg white. Except for an episode of anaphylaxis after bread ingestion containing 0.3 g of egg at 8 years of age, episodes of itching and abdominal symptoms were not noted without egg ingestion.

A telangiectatic macule was noted on her buttock at 3 months of age. As she grew older, several macular lesions developed throughout her body. At 3 years of age, new skin lesions were noted on her forehead and left-hand dorsum. At 10 years of age, physical examination revealed telangiectatic maculae on her cheeks, buttock, forehead, left-hand dorsum, and fifth finger (Fig. 1d–g), without Darier's sign (an urticarial reaction induced by skin rubbing). There was no lymphadenopathy or hepatosplenomegaly. Contrast-enhanced abdominal computed tomography revealed no signs of hemangioma or organomegaly. Except for serum total IgE levels of 289 IU/mL and antigen-specific IgEs (egg yolk, 16.60 kUa/L; egg white, 32.70 kUa/L), blood examination was otherwise unremarkable. Skin biopsy of the left finger lesion revealed mast-cell accumulation around the dilated capillaries of the upper dermis, leading to the diagnosis of TMEP (Fig. 1a–c). After OFC with 0.5 g of boiled egg yolk, she had no skin symptoms but complained of abdominal pain, suggesting a lower elicitation threshold. Considering the risk of allergic symptoms, oral immunotherapy was not introduced, and complete elimination of egg products was continued.

Pediatric mastocytosis is generally limited to the skin and tends to regress spontaneously in adolescence; however, it may progress to systemic disease in 2% of cases.³ Although Darier's sign is pathognomonic for cutaneous mastocytosis, it is usually negative in TMEP, resulting in a delayed diagnosis.¹ The frequency of food hypersensitivity in patients with mastocytosis is comparable with that in the general population, whereas food-induced anaphylaxis in patients with mastocytosis is more severe.² Moreover, the prevalence of anaphylaxis is higher in patients with mastocytosis than in the general pediatric population.⁴ The extent and density of skin lesions as well as higher serum tryptase values were identified as risk factors for anaphylaxis. Furthermore, diffused cutaneous mastocytosis and blistering episodes are risk factors for severe anaphylaxis.⁵ Infantile-onset egg allergy is outgrown with age in general, and this was also observed in the case discussed in this paper. Although the patient's egg allergy might have worsened because of complete elimination of egg products after 8 years

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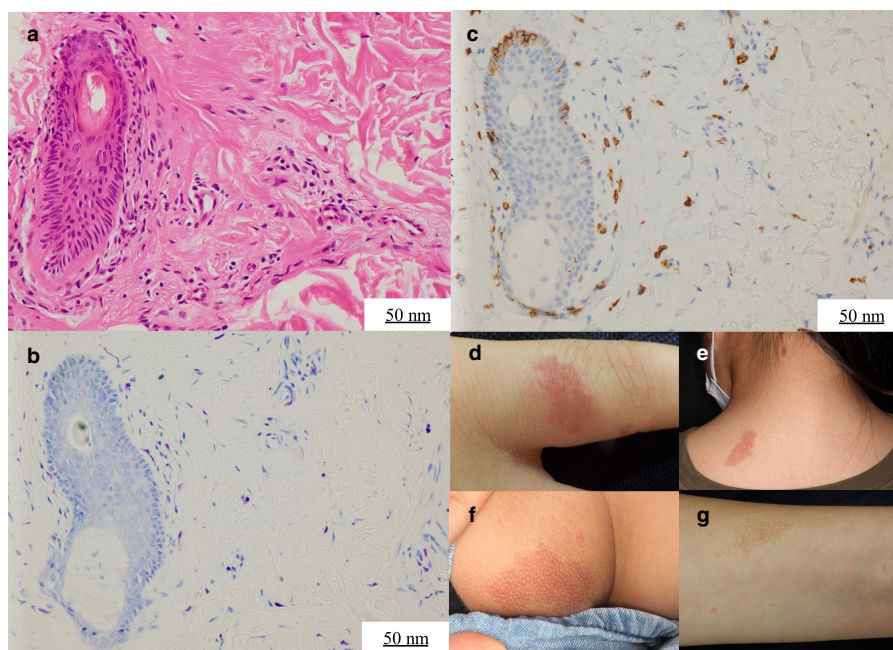


Fig. 1 Histological analysis of a telangiectasia macularis eruptiva perstans lesion. Dilated capillaries in the upper dermis with an accumulation of mast cells are visualized using hematoxylin-eosin stain (a) and toluidine blue (b). Mast cells are identified by immunohistochemistry with the anti-c-KIT antibody (c). Telangiectatic erythematous macules are distributed on the fifth finger (d), the nucha (e), the buttock (f), and the left forearm (g).

of age, the progression of skin lesions in TMEP may have lowered the elicitation threshold for allergic symptoms.

As far as we are aware, there are no case reports of TMEP associated with food allergy. However, mastocytosis should be considered in patients with persistent or exacerbated food allergies presenting with skin lesions. A skin biopsy may be helpful for diagnosing cutaneous mastocytosis in patients negative for Darier's sign. Indications for OFC and oral immunotherapy should be carefully assessed in cases of increasing cutaneous mastocytosis.

Disclosure

The authors declare no conflict of interest.

Author contributions

N.I. wrote the manuscript. M.Y. and Y.O. reviewed and supervised the manuscript. N.I., M.Y., N.O., M.H., and Y.O. treated the patient. All authors read and approved the final manuscript.

Ethical considerations

Written informed consent to publish this report was obtained from the patient's parent.

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