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CASE REPORT

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Coexistence of Frey Syndrome and Cutaneous Mastocytoma

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ABSTRACT

Auriculotemporal or Frey syndrome in children is rare and often misdiagnosed as food allergy. Here, we report a case of Frey syndrome and coincidental cutaneous mastocytoma in a 6-month-old boy. In view of the benign nature of this syndrome, no therapy is recommended. This rare disease should be kept in mind in order to prevent unnecessary diagnostic tests and elimination diets.

Keywords: Food allergy, Frey syndrome, Mastocytoma

INTRODUCTION

Frey syndrome, otherwise known as the auriculotemporal syndrome, is characterized by unilateral or, rarely, bilateral recurrent episodes of gustatory flushing and sweating along the distribution of the auriculotemporal nerve (ATN). It is relatively common in adults following injury to or surgery on the ATN. However, the condition has rarely been reported in infants and treatment is not usually recommended given the possibility of spontaneous resolution (1). Here, we report a case of Frey syndrome and coincidental cutaneous mastocytoma in a 6-month-old boy.

CASE

A 6-month-old boy presented with a 3-month history of recurrent flushing on his face after feeding. The patient had been born at term via cesarean section. Breast milk and infant formula had been given up to two months of age. Afterwards, the baby consumed either amino acid-based or extensively hydrolyzed formula due to a misdiagnosis of milk allergy. The symptoms occurred a few minutes after feeding with the bottle and disappeared within ten minutes. The erythema extended from the edge of his nose towards the tragus and forehead bilaterally without coexistent sweating, pruritus, swelling, respiratory, or gastrointestinal symptoms. Complementary feeding was introduced at about 5 months of age. The episodes were also observed a few minutes after chewing carrots and fruits and then resolved spontaneously within 10 to 15 minutes. He had no history of surgery or postnatal trauma.

Physical examination was normal, and the laboratory studies revealed peripheral blood leukocyte count 7600/mm^3; hemoglobin 12.8 g/dl; platelet count 290000/mm^3; C-reactive protein (CRP) level 0.33 mg/L; and erythrocyte sedimentation rate 2 mm/h. The tryptase level was 2.86 ng/ml. Renal and hepatic function tests were normal. Skin prick tests for common food allergens (cow's milk, egg, nuts, wheat, sesame, soya, fish, and legumes) were negative and specific IgE levels for cow's milk and egg white were within normal limits. The open challenge test with formula resulted in bilateral facial flushing that extended from the cheek to the forehead, and which resolved within 10 minutes (Figure 1A, B). The diagnosis of Frey syndrome was confirmed based on the patient's history and physical examination. In one of his follow-up visits, his mother showed a solid lesion that was slightly raised on his leg. Gentle friction of the lesion caused erythema and swelling, signifying a positive Darier's sign (Figure 2A, B). The tryptase level was doubled at that time. The patient exhibited normal peripheral blood smear and did not have

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hepatomegaly, splenomegaly or lymphadenopathy, and was therefore diagnosed as cutaneous mastocytoma. On the basis of its characteristic clinical features, skin biopsy was not performed. Informed consent was received from the family for using the patient's medical records and photos.

DISCUSSION

Frey syndrome was first described by Lucie Frey in 1923. It is defined by gustatory flushing and sweating in the preauricular area following salivary gland surgery and trauma (2). The pathophysiology of the syndrome has not been completely understood, but the most accepted mechanism is aberrant reinnervation of postganglionic parasympathetic fibers from the auriculotemporal nerve that have been injured (3). The syndrome has been reported in infants as a sequel to the perinatal birth trauma of the auriculotemporal nerve by forceps-assisted delivery. The aberrant parasympathetic fibers are stimulated during the chewing of foods, resulting in pathological vasodilatation (4). In cases where no trauma injury is apparent in the area of the parotid gland, the most valid mechanism is an anomalous congenital nerve connection. Our patient had no history of trauma. Idiopathic Frey syndrome is quite rare in childhood (5).

Frey syndrome usually manifests in early infancy with the introduction of solid foods (5,6). Chewing generates a stronger stimulation of the parotid glands. The erythema typically begins shortly after mastication of food and lasts for 15 to 45 minites and tends to be unilateral (7). In a study of forty-eight children with Frey syndrome, the erythema was unilateral in 73% and bilateral 27% of the cases (8). Since symptoms develop immediately after eating and the erythema is bilateral, food allergy was initially considered in our patient. Many cases of Frey syndrome in childhood are often falsely diagnosed as food allergy. Food allergies

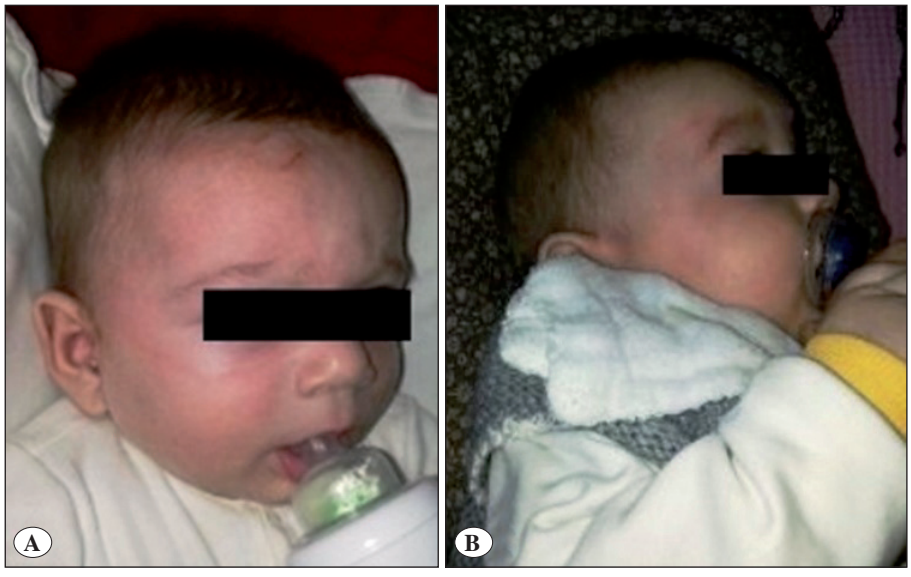

Figure 1. A, B) Bilateral facial flushing extending from cheek to forehead during open challenge test with formula.

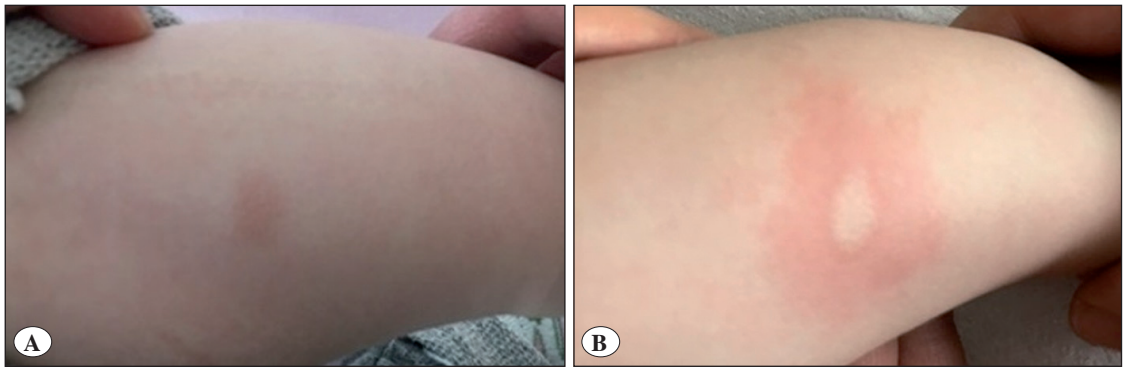

Figure 2.
A, B) Cutaneous mastocytoma, Positive Darier's sign.

are more likely to involve the face and other parts of the body and to be accompanied by itching, angioedema and respiratory or gastrointestinal symptoms (9).

Several treatment modalities have been tried in adults with Frey syndrome including anticholinergic drugs and botulinum toxin to the ATN but most of these have been proven to be ineffective (10). None of these treatments is recommended in children. Furthermore, the symptom severity tends to diminish with advancing age (7,11). In view of the benign nature of this syndrome, no therapy was suggested to our patient.

Another important finding in our case was cutaneous mastocytoma. Mastocytosis is characterized by mast cell accumulation. It is divided into two groups as cutaneous and systemic mastocytosis. This rare disorder generally appears during the first year of life. Most cases are limited to the skin (12). Mechanical irritation of lesions or using some drugs leading to the release of mast cell mediators result in localized or generalized urticaria (13). This coincidental finding in our patient might confuse the diagnosis but the clinical presentation was typical for Frey syndrome.

Here we describe a case of Frey syndrome and cutaneous mastocytoma. To our knowledge, these rare diseases appearing in the same patient has not been reported in the literature so far. Frey syndrome is self-limiting and, it does not require treatment. The diagnosis is based on a history of erythema that typically begins shortly after mastication of food in addition to careful observation of the patient during feeding. Awareness of this condition is of paramount importance for avoiding unnecessary procedures and elimination diets.

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DISCLOSURE

The authors declare no conflict of interest.

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