

Auriculotemporal's syndrome in three siblings and literature review

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Abstract

The skin is one of the target organs that are most often involved in food hypersensitivity reactions. Clinical manifestations of food hypersensitivity in the skin range from symptoms of atopic dermatitis, urticaria, angioedema to dermatitis herpetiformis, and a masquerader of food allergy, the Frey's syndrome. All present with typical skin manifestations of their foodallergic reactions. Three cases of Frey's syndrome (auriculotemporal) are presented in three children from the same family: two girls and one boy. The patients presented with a history of facial erythema in cheeks after eating different foods. Food radioallergosorbant (RAST) and percutaneous skin testing were, both, negative. Skin rash was reproduced in the clinic after oral challenge.

Introduction

Adverse food reactions are common complaints to the allergist. The diagnosis of food allergy relies on a diagnostic approach that begins with a complete history, physical examination, skin puncture, and/or specific IgE invitro testing, diet diaries, elimination of specific diets, and occasionally oral challenges [1]. When there is a discrepancy in the history, examination, testing procedures, or failure to improve with eliminationdiets, alternative nonIgE-mediated adverse reactions, not related to a specific food, need to be considered.

Auriculotemporal syndrome (Frey syndrome) is manifested as immediate unilateral or, rarely, bilateral facial flushing, sweating, or both, localized to the distribution of the auriculotemporal nerve, in response to gustatory and, occasionally, tactile stimuli [2,3]. It is not uncommon in adults and occurs as a result of surgical injury or trauma to the parotid gland. In children, the flushing typically begins a few seconds after eating and resolves approximately 30 to 60 minutes later. It is not uncommon for the auriculotemporal syndrome to be misdiagnosed as a food allergy. Here we are reporting a case of three siblings with auriculotemporal's (Frey) Syndrome. Cases

Three siblings, the first child is five years old girl, the second child is three years old girl and the third child is ten months old boy. The children were brought to the allergy clinic at Abha Medical cosultative Institute, Saudi Arabia with a history of facial erythema in cheeks after eating fruits such as orange, banana , lemon and apple. The erythema appeared immediately on mastication and disappeared within forty minutes. The erythema was extending from tragus of the ear to the lower edge of the right side of the lip. The rash was macular and not well defined. There was no pruritis, swelling, sweating or pain. The children do not have any systemic symptoms such as nausea, vomiting, cough, wheezing, stridor , throat tightness, shortness of breath or lip swelling.

Fig. 1: The three year old girl had facial erythema five minutes after oral challenge



[\(For larger image, click here\)](#)

Fig. 2: [The patient after forty minutes, erythema disappeared](#)



[\(For larger image, click here\)](#)

[Skin rash in the ten month old boy started after introducing solid food, mainly rice cereals and fruit cereals and did not follow ingestion of milk or other liquids. The rash will usually appear within few minutes of eating the cereal. Family history reveals that both mother and father have history of asthma but there were no history of eczema, food allergy, drug allergy or allergic rhinitis. Mother gave history of forceps delivery in the ten month old boy and the three years old girl. Skin test to foods such as nuts, lemon, strawberry and egg and common inhalants such dust mite, molds, pollens and animal dander were all negative. Food radioallergosorbant testing \(RAST\) was also negative. However, oral challenge with apple juice in the clinic reproduced the striking, linear erythematous flushing extending from the angle of the jaw to the corner of the mouth in all three children. The flushing was evident within ten minutes of the oral challenge test and resolved within forty minutes as shown in the photos for the three years old girl.](#)

Discussion

The pediatrician is faced with evaluating different varieties of skin rashes, a subset of which may be induced by food allergy. A complete medical history is needed to obtain not only the general medical history but also any pertinent details concerning the dietary history and any acute reactions (hives, asthma, atopic dermatitis, linear erythema, etc) to particular food ingestion [4]. A small number of foods account for >90% of the reactions [5,6,7]. In children, the most common foods that cause reactions are eggs, milk, peanut, soy, wheat, tree nuts, and fish. With two thirds of the children are reactive to eggs.⁸

A general approach is to screen children with possible allergy to: eggs, milk, peanut, soy, and wheat and, if indicated, fish and tree nuts (walnut, cashew, and pecan) by using the prick skin tests (PSTs) or RAST [9-11]. Other foods suggested by history may also be tested. If there is a significant history of food related symptoms and the tests for specific IgE to that food are positive, then the best initial treatment would be the elimination of that suspected food from the diet. The typical symptoms of immunemediated allergic food reactions are variable but may include gastrointestinal-, skin, or respiratory reactions which may be exclusively immunoglobulin (IgE)-mediated, partial IgE-mediated, or exclusively cell-mediated [12]. Cutaneous hypersensitivity disorders can also be classified as IgE-, partially IgE, or non IgE mediated. Acute urticaria and angioedema are among the most common cutaneous symptoms of IgE-mediated disorders. The onset of symptoms can be rapid, occurring within minutes of ingestion and resolve spontaneously or with treatment.

For diagnostic purposes, it is instructive to consider the prevalence of food allergy as a cause of specific disorders. For example, food allergy accounts for 20% of acute urticaria [13,14]. It is present in 37% of children with moderate to severe atopic dermatitis [9,15] and approximately 5% with atopic asthma [16] and it is the most frequent cause of anaphylaxis outside the hospital setting [17-20]. Therefore, these allergic specific disorders were considered initially in the differential diagnosis of our patients. Atopic dermatitis is a form of eczema characterized by extreme pruritis, a typical distribution of facial rash with extensor surfaces involvement, chronic relapsing course, and association with asthma or allergic rhinitis [21]. The skin rash in these three siblings was dissimilar from eczema in that it lacked oedema, xerosis, itching, and vesiculation. The rash lacks the anatomical distribution of eczema such as involvement of extensors or flexural aspects of extremities.

Urticaria is a common skin reaction that occurs at some time in the life of approximately 15% to 20% of the population [22]. It is characterized by transient erythematous, well-demarcated, raised skin lesions that may exhibit central clearing and that are usually intensely pruritic. This was not the case in our patients. Dermatitis herpetiformis is a chronic papulovesicular skin disorder which is associated with blister formation and this type of rash was not found in these three patients.

Our patients underwent for food radioallergosorbant and percutaneous skin testing and both were negative. We subsequently performed oral challenge test in the clinic with apple juice. This test showed linear erythema in all three siblings. We concluded with the above history, physical examination, negative investigations and positive oral challenge test that these three siblings suffer from Frey syndrome, also known as auriculotemporal nerve syndrome or gustatory flushing. This syndrome usually manifests as an immediate unilateral erythematous flushing and sweating over the distribution of the auriculotemporal nerve in response to gustatory or, less commonly, tactile stimuli. This observation was initially published by Frey [23] in 1923. He reported facial flushing in a patient who had suffered a gunshot wound to the face in the region of the parotid gland. In adults, the syndrome is usually a complication of surgery, trauma, disease of the parotid gland, or dorsal sympathectomy [24]. The symptoms usually include gustatory sweating in the same skin distribution. However, in children "congenital" auriculotemporal syndrome is considered a rare condition with only few described cases [24]. In reviewing the published reports, although virtually any food could elicit the rash, the most common foods reported were tangy, spicy, tart, heavily seasoned, salty, or fruity. In one patient, red foods or red candy appeared to be the most common triggers. Neither viewing or thinking about the food nor nasogastric administration of food does elicit the flushing. The onset of flushing is immediate and resolves over 30 to 90 minutes. Flushing is the primary sign in children; however, one case [19] of gustatory sweating without flushing in a teenager has been reported. The misinterpretation as a food allergy is a common pattern in most of these cases. Features that differentiate auriculotemporal nerve syndrome from food allergy or intolerance include: symptoms occurring with multiple foods, flushing, or, rarely, sweating in the same facial distribution, lack of response to antihistamines, and lack of other symptoms typical of food allergy such as urticaria, angioedema, pruritus, or gastrointestinal symptoms.

An understanding the pathophysiology of Frey syndrome requires review of the anatomy and function of the facial nerve branches. The auriculotemporal nerve is a branch of the mandibular nerve that carries sensory innervations to the skin, parasympathetic and sympathetic fibers to the parotid gland, and sympathetic fibers to the sweat glands and subcutaneous arterioles. The most frequent proposed mechanism of Frey syndrome is a misdirection of the parasympathetic fibers during healing after trauma where these fibers regenerate along the sympathetic pathway.

Mastication and salivation that result in stimulation of the parasympathetic nerve fibers that regenerated toward cutaneous blood vessels accounts for the erythema, whereas regeneration toward eccrine (sweat) glands results in sweating. The diagnosis can usually be made with a detailed history including birth history and observation with an oral challenge test. A confirmatory test if hyperhidrosis or gustatory sweating is suspected is termed the Minor test, which uses starch and iodine [25]. A tincture of iodine is applied to the skin over the affected area and allowed to dry. Next, a light layer of powdered cornstarch is applied to the area. When the patient masticates and sweating occurs, the cornstarch will turn black as it reacts to both sweat and the iodine. The rash of Frey syndrome is dissimilar to urticaria in the following ways: 1) it is nonpruritic, a macular, and does not coalesce, 2) is not affected by antihistamine therapy, 3) occurs repeatedly in the same distribution without the typical waxing/waning course, 4) may have sweating over the area, and 5) occurs with multiple unrelated foods. The treatment for pediatric auriculotemporal syndrome is usually unnecessary, as gradual reduction and even resolution of symptoms can occur [26]. The natural history of this syndrome in children is such that symptoms may spontaneously resolve over a period of several years. In general, the course is benign and nonprogressive [27]. Several treatments have been tried in adults with severe Frey syndrome including topical or systemic anticholinergic agents to diminish sweat gland function [28], topical application of 20% aluminium chloride or scopolamine cream, and local injections of alcohol [29]. Surgical procedures in the middle ear to transect the parasympathetic fibers including resection of the chorda tympani nerve are also moderately effective in adults [29]. These modalities have met with limited success and carry a risk of adverse reactions. Surgical treatment in children is therefore not recommended.

Conclusion and recommendation

In the habitual clinical practice, a great number of adverse reactions to foods may be misinterpreted as allergic reactions. This can lead to the introduction of inadequate elimination diets in children. The evolution of auriculotemporal syndrome in children is usually benign and in general it does not need treatment [30]. The knowledge of this syndrome will avoid starting several investigations in children that won't help in the diagnosis or they will mean the imposition of wrong diets of elimination. To confirm the diagnosis, simply feed the child and observe for flushing. The most beneficial approach is to provide reassurance and a clear explanation of the causes to the patient and family. We recommend that the diagnostic evaluation of food allergy to be reviewed with a careful history and physical examination in the context of a good understanding of the features of specific allergic disorders and this is the most important step toward accurate diagnosis.

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