Auriculotemporal Syndrome in Childhood

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Sir,

Because of the broad variability of clinical manifestations, paediatric dermatologists and allergologists are sometimes confronted with diagnostic difficulties when assessing food-associated reactions of the skin in children, particularly if allergological tests do not reveal any pathological results and specific elimination diets are ineffective. In these cases, children can suffer from rare diseases mimicking the cutaneous signs of food allergy.

One of these diseases is the auriculotemporal syndrome, also known as Frey's disease. Whereas it is more frequently observed in adults, e.g. after surgery to the parotid or submandibular glands, its overall occurrence in children has to be considered rare. The disease presents with a well-demarcated facial erythema and hyperhidrosis, which can be unilateral or bilateral, and, after a while, usually resolves spontaneously (3–5).

CASE REPORT

We report the case of a female patient, born by forceps delivery, who was exclusively breast-fed and did not reveal any skin symptoms during the first months of life. At the age of 6 months, however, her parents noted the development of a non-pruriginous, sharply delineated erythema on the left cheek and temporal region shortly after ingestion of different kinds of solid and liquid foods (Fig. 1). Symptoms occurred within a few seconds of mastication and spontaneously resolved after approximately 30 min without residues. Clinical examination did not reveal any signs of atopic eczema, seborrhoeic dermatitis or other skin diseases. Atopic family history was negative and no rhinoconjunctival, pulmonary or gastrointestinal signs were observed.

Under the suspicion of food allergy, previous allergological *in vitro* and *in vivo* diagnostic procedures including an ImmunoCAP® assay (Pharmacia, Uppsala, Sweden), skin prick-test and an atopy patch-test had been performed outside our clinic and, collectively, revealed no pathological results. Still, on the recommendation of the family physician the patient had been kept on an elimination diet without cow's milk when we saw her for the first time in our division of Paediatric Dermatology.

After thoroughly examining the clinical history, we performed oral provocation tests with the incriminated food agents on different occasions and were able to repeatedly reproduce the occurrence of a unilateral, sharply demarcated, non-hyperhidrotic reddish flushing in the left auriculotemporal area of the infant in response to distinct gustatory stimuli irrespective of the kind of food given to the patient. Although the cutaneous signs intermittently diminished during clinical follow-ups, they were still visible at the age of 12 months and, to date, flushing still occurs. We made a diagnosis of auriculotemporal syndrome based on the clinical symptoms.

DISCUSSION

Allergic reactions are known to account for the majority of food-associated skin reactions in children. Thus, allergological testing frequently results in identification of the disease-eliciting agent and a subsequent therapeutic elimination diet. However, in cases such as the one presented here, these tests do not reveal any allergy, and specific diets do not lead to improvement of the clinical symptoms.

Upon examination of our patient, allergic and intolerance reactions to food could be ruled out because symptoms always occurred in the same unilateral localization following ingestion of different unrelated nutritive agents without any associated systemic allergic symptoms such as, for example, diarrhoea, angioedema or pruritus. Further differential diagnoses, such as unilateral naevus flammeus, ulerythema ophryogenes or

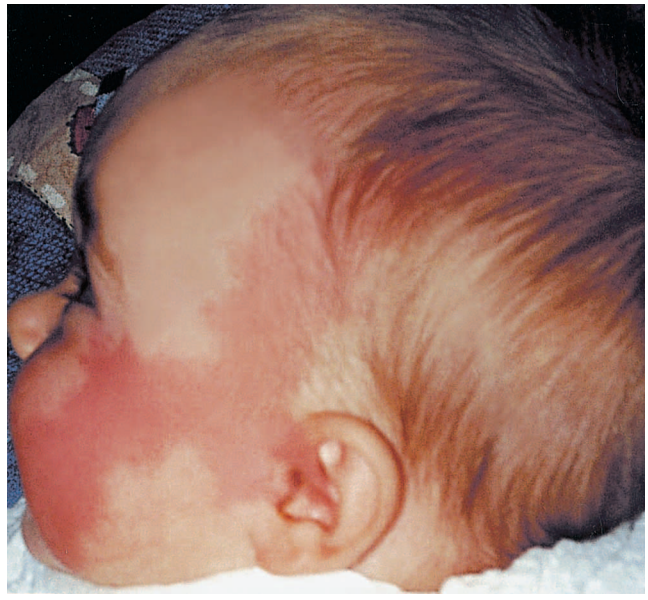

Fig. 1. Non-pruriginous, sharply delineated erythema on the left cheek and temporal region shortly after ingestion of cow's milk.

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atopic eczema, could also be ruled out clinically. After further interrogation of her parents, the mother reported that our patient had been born by forceps delivery, a procedure that potentially can lead to trauma of the parotid or submandibular glands. Taking these data into account, we established the diagnosis of auriculotemporal syndrome.

This hyperhidrotic, food-associated facial flushing in the auriculotemporal region is also known as gustatory hyperhidrosis or Frey's disease, named after the Polish neurologist Lucja Frey, the first to publish pathophysiological reflections on this phenomenon in 1923 (3). While the characteristic clinical symptoms are observed after parotid surgery in up to 98% of adult patients, the disease has only rarely been described in childhood (4, 5), depending on the population studied (6). To our knowledge, less than 40 paediatric cases have been published so far.

To date, the aetiology of Frey's syndrome is only partly understood. Surgical procedures, inflammatory reactions or injuries to the parotid gland are thought to damage parasympathetic fibres of salivary glands. During regeneration, these parasympathetic fibres are misdirected and wrongly innervate skin vessels or sweat glands that are normally innervated by sympathetic fibres. This misdirected regeneration is thought to result in a short circuit with localized sweating and auriculotemporal erythema during and after salivation (7).

Perinatal birth trauma in children, such as the forceps delivery reported in our patient, is considered a possible cause of this condition and previously has been communicated in other infants and children with Frey's syndrome (8). However, congenital cases without known traumatic aetiology have also been published and were attributed to central nervous system abnormalities or subclinical intrauterine infection (9).

Whereas in adults the symptoms of gustatory sweating and flushing are permanent if occurring after surgical procedures, these symptoms usually resolve spontaneously in children, an observation that is not understood yet and still requires investigation. Since remission is thought to occur spontaneously within the first years of life, dietary restrictions or other therapeutic measures in children are unnecessary, if not even harmful. Haxton described the case of a child who underwent cervical sympathectomy for Frey's disease and subsequently developed Horner's syndrome (10). Additionally, elimination diets without prior allergological counselling are known to distress patients and cases of malnutrition due to unjustified avoidance of cow's milk have been published repeatedly (11, 12). Botulinum toxin, an agent used in the treatment of adult patients with auriculotemporal syndrome, is routinely used in children with cerebral palsy (13). However, hyperhidrosis was not present in the case presented here and therefore the use of botulinum toxin is not indicated. Further, serious side effects and the narrow therapeutic range of this drug clearly represent a contraindication for its application in such benign and self-limiting conditions as Frey's disease.

To avoid time-consuming and unnecessary diagnostic procedures as well as potentially harmful elimination diets and therapeutic regimens it is therefore crucial to consider even rare differential diagnoses like the auriculotemporal syndrome when children reveal food-associated cutaneous symptoms.

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