Case Report

CRANIAL NERVE MISCHIEF MASQUERADING AS FOOD ALLERGY: AURICULOTEMPORAL (FREY’S) SYNDROME IN A CHILD WITH MOEBIUS SYNDROME

Claudia L Gray | MBChB, FRCPPCH, MSc, PhD, DipAllergy, DipPaedNutrition
Laura Berry | BSc (Diet), PGDip (Diet), RD

Division of Allergy, Red Cross War Memorial Children’s Hospital, Cape Town
Email | claudiagray.paediatrics@gmail.com

INTRODUCTION

The diagnosis of food allergy can be challenging and many conditions can mimic food allergy. It is imperative to differentiate true food allergy from the ‘mimics’ of food allergy, particularly in children eliminating multiple foods from their diet. This case report describes a child with features of Auriculotemporal (Frey’s) syndrome, posing as multiple food allergies. The child was known to have Moebius syndrome, a rare neurological disorder characterised by weakness or paralysis of a group of facial nerves. Auriculotemporal syndrome as a potential complication of Moebius syndrome has not been described previously.

CASE

Patient GP, 21 months of age, was diagnosed at a few months of age with Moebius syndrome, based on the clinical picture of initial feeding difficulties, lack of facial expression and typical facial features including micrognathia. At approximately 12 months of age, on second exposure to hen’s egg, he experienced almost immediate onset vomiting and floppiness. Subsequent egg-allergy testing by skin-prick test (SPT) was positive, supporting the diagnosis of true IgE-mediated reaction to egg. He also seemed to become crampy after consumption of cow’s milk-based formulas, hence cow’s milk protein was being eliminated from his diet on the advice of his clinician. His SPT to soya milk had come up as 3 mm on a previous occasion, so his clinician advised him to avoid soya, although he had never consumed it before. Distressingly for his mother, he also seemed to react to a wide variety of other foods, often those with a ‘stronger’ or slightly ‘sour’ flavour, including apples, strawberries, oranges, dried mango, sunflower oil and soya. Consumption of these foods led to immediate blotchy flushes on the side of his cheek – usually on the left side – which lasted a few minutes before resolving spontaneously, and did not seem to distress him. There were no obvious hives and no other signs of an IgE-mediated reaction on consumption of such foods. Nonetheless, all of these foods were eliminated from his diet by his mother for fear that they represented true allergies to these foods. The multiple dietary exclusions made for a limited diet and great difficulty in the preparation of toddler-type foods.

Examination revealed a 21-month-old boy with a lack of facial expression and difficulty in abducting both eyes. His skin was clear and systemic examination otherwise normal. His weight of 10.5 kg and height of 83 cm were on the 25th centile for age. Investigations showed the following:

<table>
<thead>
<tr>
<th>FOOD</th>
<th>TEST AND RESULTS</th>
</tr>
</thead>
<tbody>
<tr>
<td>Cow’s milk</td>
<td>SPT 0 mm</td>
</tr>
<tr>
<td>Egg white</td>
<td>SPT 3 mm (≥3 mm considered positive)</td>
</tr>
<tr>
<td>Fresh raw egg white</td>
<td>SPT 8 mm (positive)</td>
</tr>
<tr>
<td>Orange</td>
<td>SPT 0 mm</td>
</tr>
<tr>
<td>Strawberry</td>
<td>SPT 0 mm</td>
</tr>
<tr>
<td>Tomato</td>
<td>SPT 0 mm</td>
</tr>
<tr>
<td>Soya</td>
<td>SPT 0 mm</td>
</tr>
<tr>
<td>Sunflower seed</td>
<td>Specific IgE 0.08 kU/L (normal range &lt;0.35 kU/L)</td>
</tr>
</tbody>
</table>

An IgE-mediated egg allergy was confirmed. A baked egg challenge was passed and egg was avoided in the raw and whole forms, for six-monthly review.

There was no evidence of IgE-mediated reactivity for the other foods which produced temporary flushing, hence a diagnosis of likely Auriculotemporal Nerve Syndrome (Frey’s syndrome) was made.

This diagnosis was strengthened by the fact that the patient had involvement of multiple cranial nerves as part of Moebius syndrome, and we theorised that possible involvement of the parasympathetic fibres of the auriculotemporal nerve, which is fed by the glossopharyngeal nerve (cranial nerve IX), contributed towards symptoms.

The foods which caused temporary flushing were reintroduced into his diet successfully, with reassurance of the benign nature of symptoms, providing much-needed dietary diversity.

DISCUSSION

Moebius syndrome is a rare, non-progressive congenital neurological disorder which most often presents with a mask-like face and inability to abduct the eyes as a result of weakness or paralysis of the facial nerve (cranial nerve IX).
CASE STUDY

VII) and abducens nerve (cranial nerve VI). It occurs sporadically, although familial cases have been described. Other cranial nerves including the Vth, VIIIth, IXth, Xth, XIth and XIIth cranial nerves may be involved. One series in children with Moebius syndrome found that the glossopharyngeal nerve (IXth cranial nerve) was involved in 22 per cent of cases. Other abnormalities include underdevelopment of the pectoral muscles, defects of the limbs and dental problems. An association with food allergies or food allergy-like symptoms has not been described previously.

Auriculotemporal syndrome (Frey’s syndrome) is a rare neurological disorder resulting from damage around the parotid glands or damage to the auriculotemporal nerve. Symptoms are redness/flushing and sweating in the cheek area (usually in response to gustatory stimuli), especially when the person eats certain foods with a strong flavour which produce strong salivation. The auriculotemporal nerve arises as two roots from the posterior division of the mandibular nerve, a branch of the trigeminal nerve (cranial nerve V). It supplies somatosensory innervation to the auricle, external acoustic meatus, tympanic membrane and skin in the temporal region. It also supplies parasympathetic fibres to the parotid gland; these originate from the glossopharyngeal nerve (cranial nerve IX). The underlying theory for pathophysiology of Frey’s syndrome is that damage to the auriculotemporal nerve leads to fibres which previously innervated the parotid gland to reattach to sweat glands in the same region (see Figure 1). The result is sweating and flushing along the auriculotemporal nerve territory: the lateral cheek, medial to the ear and the frontotemporal scalp (see Figure 2). Symptoms begin shortly after chewing of food and last 15–45 minutes. These symptoms are not accompanied by itching, burning or discomfort.

In children, Frey’s syndrome is rare, but usually manifests during infancy with the introduction of solid foods. The use of forceps during a delivery is a known risk factor due to trauma to the parotid region. In our case, possible malformation of the auriculotemporal nerve as a result of Moebius syndrome is postulated. In contrast to the syndrome in adults, gustatory sweating is rarely associated with children, possibly due to immaturity of the sweat glands, and they present mainly with flushing. Because of the development of symptoms after eating, many patients are initially suspected of having a food allergy. In children, gustatory flushing is benign and non-progressive and no specific treatment is necessary. Often the flushing disappears spontaneously with time. Injections of botulinum toxin A, surgical transection of nerve fibres and application of anticholinergic ointments have been used, but the risk-versus-benefit ratio make them inappropriate choices for most children.

CONCLUSION
Auriculotemporal syndrome is a rare ‘mimic’ of food allergy caused by malfunctioning of the auriculotemporal nerve; in this case as a possible result of Moebius syndrome, which is known to involve multiple cranial nerves. Recognition is paramount to avoid mislabelling as food allergy and unnecessary dietary restrictions.

DECLARATION OF CONFLICT OF INTEREST
The author declares no conflict of interest.

This article has been peer reviewed.

REFERENCES