CASE REPORT

Frey syndrome following herpes zoster in an otherwise healthy girl

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SUMMARY

A 12-year-old girl presented with red spots appearing on the left side of her face. The girl was usually healthy and fully vaccinated, including varicella vaccination.

Six years prior to her presentation, she had suffered an episode of blister rash on the left side of her face, including lesions in the ear canal and buccal mucous membrane. A diagnosis of herpes zoster was made, and she was treated with acyclovir with complete skin recovery. A hearing examination demonstrated mild-to-moderate left neurosensory hearing loss.

Since then, she is having short episodes of redness on her face without pain or sweating at the exact distribution of the zoster blisters 6 years ago. The appearance of spots is related to sour foods, such as sour flavoured candies, yoghourt and green apples. The diagnosis of postherpetic Frey syndrome was made, and observational approach was adopted due to the benign character of symptoms.

BACKGROUND

We believe that this is a rare and interesting case. The phenomenon described in our case may be underdiagnosed and actually more common. Although Frey syndrome cases following parotid gland surgery and other trauma are relatively commonly reported, it is very rare to occur after herpetic infection. To the best of our knowledge, only two paediatric,[1 2] and one adult case,[3] of postherpetic Frey syndrome were published until now. The postprandial appearance of the phenomenon may lead to erroneous diagnosis of food allergy which in turn may lead to unnecessary tests and treatment.

CASE PRESENTATION

A 12-year-old girl presented with a periodic recurrent rash on her face.

Her mother described a red colouration appearing on the left side of her face without any specific known provocation. It was not painful but was embarrassing. She has been a healthy girl, immunised to her age, including the chickenpox vaccine.

In August 2011 (when she was 6 years old), she had an acute event of blister rash on the left side of her face, including lesions in the ear canal and buccal mucous membrane. The diagnosis of herpes zoster was made, and the girl was treated with acyclovir with complete skin recovery. A hearing examination done a few weeks later demonstrated mild-to-moderate neurosensory hearing loss on the left side. She then underwent an MRI of the brain that was found normal.

Her hearing loss is not progressing.

The girl and her parents remarked that since the herpes zoster episode, she was having short episodes of redness on her face at the exact same distribution of the original zoster blisters 6 years ago (figure 1). The redness appears unprovoked, without pain or any other sensation and without sweating. It subsides after a few minutes. After several trials, it seems that the rash appearance is related to sour foods, such as sour flavoured candies, yoghourt and green apples.

DIFFERENTIAL DIAGNOSIS

Skin flushing following food exposure can be erroneously interpreted as food allergy, the main differential diagnosis of the condition. Precise history and the induction of flushing only by visual stimulation (images of food) can help to distinguish between the phenomena. Since the neurogenic origin of the condition became clear, further differential diagnoses regarding auriculotemporal nerve injury are discussed in the 'Discussion' section.

OUTCOME AND FOLLOW-UP

In our case, considering the benign nature of the girl's symptoms, only an observational approach was adopted. The flushing episodes still appear, but are less disturbing to the girl. The patient and her parents report that after receiving an explanation and reassurance, she is free from any psychological distress and is enjoying a normal social life.

DISCUSSION

Frey syndrome, gustatory sweating and flushing of the involved skin, is usually depicted in the context of an iatrogenic sequela after a parotid gland region surgery or a trauma to this region.[4]

The condition is named after Lucja Frey, a Polish–Jewish neurologist, who described the auriculotemporal nerve anatomy and the syndrome pathophysiology in 1923.[5] The syndrome is caused by damage to the auriculotemporal nerve, a sensory nerve that contains both postganglionic sympathetic and postganglionic parasympathetic fibres. Although the condition and its management are well described in the context of parotidectomy in adults, only few cases in children and particularly following herpes zoster infection were published in the literature.[1] Recently, Shah and Asrani described a similar case in a 27-year-old man.[3] Most cases

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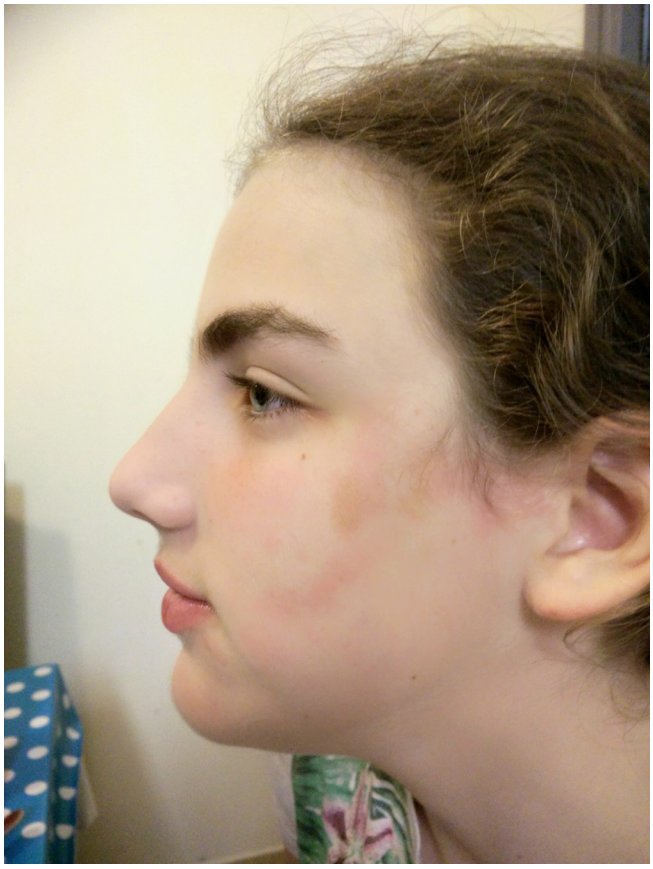

Figure 1 Unilateral flushing in the area innervated by the left auriculotemporal nerve.

described in infants and small children are associated with obstetric trauma and specifically forceps delivery.[6 7] In paediatric population, the condition was also described following trauma and neurofibroma.[8 9] Sethuraman and Mancini described two sisters with bilateral auriculotemporal syndrome, postulated a congenital aberration of the auriculotemporal nerve pathway between parasympathetic and sympathetic fibres.[10] Drummond *et al* described, in 1987, an analogous condition after herpes zoster in the third division of the trigeminal nerve.[2] In our case, skin affected by varicella zoster virus herpetic lesions in the distribution of the auriculotemporal nerve receives sympathetic innervation from the plexus surrounding the middle meningeal artery. Following damage of the sympathetic fibres at the time of herpetic invasion, development of aberrant collateral parasympathetic fibres along the degenerated sympathetic pathways leads to innervation of subcutaneous blood vessels and sweat glands, leading in turn to flushing and sweating following gustatory stimulation.[2 11 12] Only skin flushing without the sweating symptom is characteristic in children as opposed to adult patients.[6 8] It is remarkable that in the series of eight cases described by Dizon *et al*, most of the children were misdiagnosed as allergic reaction to food,[6] thus the acknowledgement with the syndrome is important for prevention of unnecessary tests.[6 7] Probably, the differentiation is especially difficult in small children. Botulinum toxin injections can provide good results in postoperative cases,[13] but to the best of our knowledge, there is no experience of such a treatment in the paediatric population.

Learning points

- The physician should inquire about the history of herpes zoster in the evaluation of unexplained flushing.
- The acknowledgement with the phenomenon can prevent unnecessary allergy workup.
- Most children with Frey's syndrome exhibit flushing without sweating.

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REFERENCES

1 Bourgeois P, Morren MA. FRey's syndrome after herpes zoster virus infection in a 2-year-old girl. *Pediatr Dermatol* 2015;32:e184–e185.
2 Drummond PD, Boyce GM, Lance JW. Postherpetic gustatory flushing and sweating. *Ann Neurol* 1987;21:559–63.
3 Shah JS, Asrani VK. POst herpetic frey's syndrome. *Ann Maxillofac Surg* 2017;7:278–81.
4 de Bree R, van der Waal I, Leemans CR. Management of frey syndrome. *Head Neck* 2007;29:773–8.
5 Frey L. Le syndrome du nerf auriculo-temporal. *Revue Neurologique* 1923;2:92–104.
6 Dizon MV, Fischer G, Jopp-McKay A, *et al*. Localized facial flushing in infancy. Auriculotemporal nerve (Frey) syndrome. *Arch Dermatol* 1997;133:1143–5.
7 Caulley L, Hong P. Pediatric auriculotemporal nerve (Frey) syndrome. *CMAJ* 2013;185:504.
8 Kaddu S, Smolle J, Komericki P, *et al*. Auriculotemporal (Frey) syndrome in late childhood: an unusual variant presenting as gustatory flushing mimicking food allergy. *Pediatr Dermatol* 2000;17:126–8.
9 Listernick R, Legius E, Charrow J. Gustatory flushing (auriculotemporal nerve syndrome) in children with neurofibromatosis type 1 and facial plexiform neurofibromas. *J Pediatr* 2011;158:1034.e1.
10 Sethuraman G, Mancini AJ. Familial auriculotemporal nerve (Frey) syndrome. *Pediatr Dermatol* 2009;26:302–5.
11 Gardner WJ, McCubbin JW. Auriculotemporal syndrome; gustatory sweating due to misdirection of regenerated nerve fibers. *J Am Med Assoc* 1956;160:272–7.
12 Murray JG, Thompson JW. Collateral sprouting in response to injury of the autonomic nervous system, and its consequences. *Br Med Bull* 1957;13:213–9.
13 Beerens AJ, Snow GB. Botulinum toxin A in the treatment of patients with Frey syndrome. *Br J Surg* 2002;89:116–9.