Idiopathic Frey’s Syndrome under the appearance of a recurrent otitis externa

S. Santa Cruz Ruiz*, A. Muñoz Herrera*, P. Santa Cruz Ruiz**, M. Gil Melcon*, A. Batuecas Caletrio*


Abstract: Frey syndrome has been observed especially in patients who have undergone a parotidectomy operation, but also in zoster herpes, in parotiditis, condyle fractures, obstetric traumatisms with forceps and in surgery of the meningoima of the cerebellopontine angle. It also appears without previous surgery, like in our case. In these circumstances it is believed that a clinical neuritis, primary or secondary to a neighbouring inflammation may cause the start of this disorder. Several treatments have been suggested which highlights the difficulty of them. The most effective one is the intradermic injection of botulinum toxin type A. It’s use in Frey’s syndrome was initiated by Drobik and Laskawi in 1995.Since then the references to its use are numerous. Nevertheless, it is a treatment which has been introduced very few times in our country.

Key words: Frey’s syndrome. Auriculotemporal syndrome. Botulinum toxin. Etiology.

INTRODUCTION

Frey’s syndrome, or auriculotemporal syndrome, was described in 1923 by the French neurologist Lucie Frey. She described a syndrome characterized by the appearance of erythema and sweating, localized at the level of the cervico-facial region, which is related to diet and makes its appearance after an ipsilateral parotid trauma. Since then it has been observed mainly in patients who have undergone a parotidectomy, but also in association with trigeminal shingles (herpes zoster), parotiditis, condyle fractures, obstetric trauma from forceps, and surgery for meningoima of the cerebellopontine angle1.

The most accepted hypothesis is that of an anomalous crossed reinnervation of the parasympathetic fibers of cranial pair IX that go up to the sweat glands of said areas. In healthy people these sweat glands normally receive sympathetic innervation, but after a parotidectomy the parasympathetic fibers that pass through the auricular temporal nerve, when sectioned and subsequently regenerated, reach the sweat glands as well as the veins of the skin. The neurotransmitter that works in the parasympathetic fibers is acetylcholine.

The origin of these parasympathetic fibers is complex: they start off from the inferior salivary bulbus nucleus, situated under ventricle IV. These fibers continue along cranial pair IX, from where the Jacobsen nerve originates, which crosses the promontory in the middle ear. One of its branches, the minor superficial petrous nerve, goes as far as the otic ganglion, origin of the auriculotemporal nerve, until finally it reaches the parotid gland, stimulating salivary secretion to different food-related stimuli.

According to estimations, this syndrome occurs in 20% of children and 65.9% of adults who have undergone a parotidectomy1. However, the Minor - or starch-iodine - tests are positive in almost 100% of cases. This test consists of painting the skin that shows the symptoms with iodine, allowing it to dry, then applying a fine layer of starch on top and stimulate salivary secretion that will cause sweating. The starch is mixed with the iodine, turning it into a dark color of convergent form.

Amongst the many treatments proposed we should highlight the external application of:

- Roll-on deodorant
- Glycopyrrolate
- Atropine
- 3% Scopolamine cream.
- Sectioning of the Jacobson nerve, by means of tympanotomy.
- Insertion of a flap of sternocleidomastoid muscle between the skin and parotid layer.
- Insertion of fascia lata or other autologic or artificial tissues like Silastic.
- Removal and ligation of the auriculotemporal nerve; alcoholization of the superior cervical ganglion; otic...
ganglion; lingual nerve; tympanic plexus; tympanic cord, etc.

- Intradermic botulinum toxin type A injection.

Botulinum toxin is one of the seven neuro-toxins produced by the anaerobic bacteria Clostridium botulinum. It is fixed to the presynaptic receptors of the cholinergic nerve endings, inhibiting the freedom of the acetylcholine, which involves a blockage of the motor plates and a fall in the cholinergic receptors at this level3,4.

The use of this treatment in sweating pathologies began with axillary and palmar hyperhidrosis5. Drobik and Laskawi administered it for Frey’s syndrome in 19956. Since then there have been numerous references to its effectiveness. However, it is a treatment rarely described in our country as we have been able to prove in a review carried out of the existing publications on PubMed.

CLINICAL CASE

Medical history

A male of 68 years who had attended our consultancy since December 1999 for problems in the left ear that had lasted three years, with aqueous otorrhea during ingestion and discomfort related to cold temperatures. He did not have a previous history of otitis, or itching, or manipulation in the external auditory canal. His hearing was normal.

Personal history

The only antecedent which stood out was insufficient nasal ventilation related to the collapse of the cartilaginous pyramid after Killian’s septal surgery.

Physical examination

The otoscopy showed a small osteoma on the right side situated at 5 o’clock and on the left side a wide EAC. The skin was neither pitted nor scaly. A small quantity of thick white otorrhea and a slightly swollen tympanic membrane were observed.

Initial diagnosis

We diagnosed chronic external otitis, probably infectious, secondary to a Klebsiella oxytoca and sensitive to external ciprofloxacin.

The patient showed partial improvement from the treatment, although the otorrhea persisted there was no oedema of the skin in the EAC.

In the face of the persistence of the symptoms, new samples were taken for a microbiological study (many were negative, others positive for Klebsiella Pneumoniae, Aspergillus flavus) and different external antibiotics were tested; neomycin, polymyxin A, bifenazol, 5% acetic acid with flucnazole by general administration. There was only a partial improvement while the treatment lasted, and the symptoms subsequently reappeared.

In the differential diagnosis we considered the possibility of a salivary fistula between the parotid gland and the EAC, although neither a microscope nor a thorough endoscopy showed any fistula in the EAC, tympanic bridle, etc, ….which was ruled out with a normal parotid sialography.

Secretion increased during chewing, and the level of amylase, ions and proteins was determined from a sample in order to compare them with the levels in the blood. The amylase in the blood was 195 U/L (1-100) and 97 U/L in the exudation, a result that persuaded us to carry out an operation with local anesthetic to explore the contact space of the parotid gland and the cartilaginous portion of the EAC. We did not find a fistular tract, so we inserted a silastic plate in order to place a barrier between both structures.

The secretion came to an end over a period of two months, with the subsequent re-appearance of the same symptoms.

At this stage of development, the patient suffered an intense depression which was related to the persistence of the otic problems and the failure of the different treatments.

In the differential diagnosis we considered the possibility of Frey’s syndrome. For this reason we performed the Minor test which was clearly positive (Figure 1).

Treatment

We started treatment with injections of congealed botulinum type A toxin, reconstituted with physiological serum, infiltrating 2.5 UI into each centimeter square of skin up to a total dosage per session of between 5 and 7.5 U. The intradermic injection was carried out using an intradermic monject syringe 29G without any type of anesthetic (Figure 2). After five sessions, with intervals of two months, we obtained a clear reduction of the sweating until it was imperceptible.

DISCUSSION

Frey’s syndrome is generally associated with a parotidectomy and is explained by the hypothesis of the aberrant regeneration of the parotid parasympathetic fibers. However, this disorder can also appear without prior surgery, as in the case we are presenting. In these circumstances, the etiology can correspond to a subclinical neuritis, primary or secondary to a neighbouring infection, as was the origin with this disorder.
In our case, as was already known\textsuperscript{7,8}, the technique of placing a barrier between the parotid gland and the skin failed.

The benefit of the application of different anticholinergic agents in the treatment of the hyperhidrosis\textsuperscript{9} is well demonstrated, showing an effectiveness of 60-70\%. Its disadvantages are: its effect disappearing after 48 hours, numerous contraindications and multiple side effects.

On the other hand, the advantage of botulinum type A toxin is that it is a non-surgical treatment, it can be carried out on an outpatient basis, and the symptoms improve or disappear in 2-5 days, with scarce and mild secondary effects. In addition, its definitive effectiveness is nearly 100%.

References