Auriculotemporal nerve syndrome in association with congenital haemangiopericytoma: a case report

**ABSTRACT**

**Background** Auriculotemporal nerve syndrome is characterised by recurrent episodes of facial gustatory flushing and/or sweating along the cutaneous distribution of the auriculotemporal nerve. The condition is rare in children and is normally a sequel of perinatal birth trauma. We report a case of a sixteen-month-old boy referred by paediatric oncology with recurrent, unilateral facial flushing of the left cheek which had been present for 2 months. The flushing only occurred during mastication. The patient had also received treatment for a rare vascular tumour, congenital haemangiopericytoma, of the left cheek and parotid region. The possible association between auriculotemporal nerve syndrome and congenital haemangiopericytoma is discussed. Knowledge of the presentation, aetiology and management of Auriculotemporal Nerve Syndrome can provide much needed reassurance to those suffering with this condition.

**Keywords:** Auriculotemporal Nerve Syndrome; Frey’s Syndrome; Haemangiopericytoma

**Introduction**

Auriculotemporal nerve (ATN) syndrome or Frey’s syndrome is characterised by unilateral recurrent facial sweating or flushing that occurs when salivation is stimulated. The symptoms occur in the cutaneous distribution of the auriculotemporal nerve, a branch of the mandibular nerve. The condition gained its name following an illustrative article by Frey’s in 1923 [Frey, 1923], however the first reported case of Frey syndrome should be attributed to Baillarger in 1853, as quoted by Dulguerov et al. [1999].

ATN syndrome is relatively common in adults as a complication of parotidectomy and may follow other surgical, traumatic, and inflammatory injuries of the parotid and submandibular glands [Malatskey et al., 2002]. Rarely, ATN syndrome has been reported in infancy as a sequel of perinatal birth trauma resulting from assisted forceps delivery [Dizon et al., 1997]. We recently observed an unusual case of ATN syndrome in a 16-month-old child diagnosed with congenital haemangiopericytoma (HPC) extending from the parotid gland to the skull base.

HPC is an uncommon, vascular tumour that originates from small pericapillary spindle-shaped cells known as pericytes [Stout and Murray, 1942]. The HPC is mainly a tumour of adulthood and occurs in lower extremities although 26% occur in the head and neck region mostly from the soft tissues of the scalp, face and nasal cavities [Enzinger and Smith, 1976; Billings et al., 2000].

The aetiology of HPC is unknown; however the tumour has been linked to prolonged steroid use, hormonal disturbances and trauma [McM aster et al., 1975]. Diagnosis of HPC of the head and neck region can be challenging with the differential diagnosis including Chondrosarcomas, Ewing’s sarcoma, Osteosarcoma, Juvenile haemangioma and Leiomyosarcoma. Definitive diagnosis of HPC relies on histopathological assessment [Carvalho et al., 2004].

**Case report**

A sixteen-month-old boy was referred to the Paediatric Dentistry Unit of the Charles Clifford Dental Hospital by the paediatric oncology team. The patient presented with a 2 month history of recurrent, unilateral facial flushing of the left cheek. The flushing only occurred during mastication, (Fig. 1, 2) and appeared to coincide with the eruption of the first primary molar teeth and the introduction of solid foods. There was no associated discomfort, swelling or perspiration and no suggestion of food allergies. The patient was an only child and was born at 41 weeks by caesarean section. There was no relevant family history.

Extra-oral examination revealed a biopsy scar posterior to the left ear. There was no facial flushing, swelling or lymphadenopathy (Fig. 1). It was observed that during mastication (the patient was given a piece of fruit), an instant and florid flushing of the left cheek occurred, extending from the left pre-auricular area to the corner of the mouth (Fig. 2). On intra-oral examination eleven primary teeth were present and the oral soft tissues were normal. Of significance, the patient had been previously diagnosed with congenital HPC, a rare, benign vascular tumour. The lesion had presented on the left cheek and parotid region (Fig. 3), extending to skull base on MRI (Fig. 4). Diagnosis was confirmed by incisional biopsy at 10 weeks of age (Fig. 5). Peri-operatively the patient required platelet transfusions due to thrombocytopenia secondary to platelet consumption (Kassabach-Merritt Syndrome). Subsequently the tumour was managed with chemotherapy over 12 months. The regimen included vincristine, actinomycin, and cyclophosphamide for five months followed by vincristine and actinomycin for seven months. Surgery and radiotherapy were not employed. The lesion responded well to chemotherapy and a follow-up MRI at 19 months excluded tumour recurrence (Fig. 6).
Discussion

Kauffman and Stout in 1960 recognised that congenital/infantile HPC, which accounts for 10% of all HPCs, differs from its adult counterpart in behaviour and response to therapy [Kauffman and Stout, 1960]. Congenital HPC is reported to follow a benign clinical course although metastases can occur rarely [Hoey et al., 1998]. A review of the literature identified 84 reported cases of congenital HPC [Hoey et al., 1998]. Our literature review revealed a further 17 cases including our case bringing the total to 102.

The mainstay of treatment for paediatric HPC is surgical resection where possible [Rodriguez-Galindo et al., 2000]. The role of adjuvant chemotherapy has yet to be defined due to the rarity of this condition. In unresectable cases of HPC, chemotherapy alone has been successfully employed with chemotherapeutic regimens involving the use of vincristine, doxorubicin, actinomycin and cyclophosphamide [Craven et al., 1992]. Although radiotherapy has a role in treatment, the long-term sequelae in this age group should be strongly considered [Rodriguez-Galindo et al., 2000].

In this case, due to the early age at presentation and the extent of the lesion, chemotherapy was the only treatment modality used. The successful clinical response reinforces the high chemoresponsiveness of infantile HPC reported previously [Rodriguez-Galindo et al., 2000; Yamanishi et al., 2007].

To our knowledge, this is the first report of congenital HPC of the parotid gland with ATN syndrome. In our patient the occurrence of the ATN syndrome can be
explained by several mechanisms. This includes direct trauma to the ATN due to tumour invasion, which seems plausible considering the extent of the lesion which involved the left parotid gland extending to the skull base with erosion and extension into the left mastoid and adjacent muscles. Direct surgical trauma could also have occurred during the open biopsy or ensuing oedema. Of note, the specimen was small in size and there were no nerve fibres evident on histological analysis. Finally, direct pressure on the ATN and eventual malconduction of stimuli could arise as a result of scar tissue formation during healing [Kaddu et al., 2000]. Interestingly, the occurrence of symptoms coincided with eruption of first deciduous molars and hence an increase in the patient’s chewing capability. It is therefore possible that ATN syndrome had developed much earlier but only became evident when the patient’s diet changed to more solid food consumption.

Currently the standard treatment for ANT syndrome is intradermal injection of botulinum toxin [Guntinas-Lichius, 2002; Pomprasit and Chintrakarn, 2007]. With this therapy symptoms can be abolished for up to 1.5 years [Guntinas-Lichius 2003]. Other modalities include surgery and the use of topical anticholinergic medications. The possible use of botulinum toxin in the future has been discussed with the parents. However, as the condition in children often resolves spontaneously [Dizon et al., 1997; Kaddu et al., 2000], treatment could be deferred until the patient becomes aware of his symptoms.

Conclusion

This is the first case report of ATN syndrome in association with Haemangiopericytoma. It is important for paediatric dentists to be aware of ATN syndrome and its possible presentation in patients with parotid tumours in order to effectively manage and reassure patients with this condition.

Acknowledgement

The authors would like to thank Dr. M. Al-Adnani, Consultant Paediatric Histopathologist, for his assistance with this case report.

References


