DOI: 10.1111/j.1365-263X.2008.00950.x

# Glial choristoma of the tongue: report of a case and clinico-pathological features

**DAVID JAMES BALDWIN[1], THAYALAN KANDIAH[1], AMRITA JAY[2] & FERRANTI WONG[3]**

[1]Paediatric Dentistry, Dental Institute, The Royal London Hospital, New Road, Whitechapel, London E1 1BB, UK, [2]Oral & Maxillofacial and ENT Pathology, Histopathology, University College Hospital, Rockerfeller Building, London WC1E 6JJ, UK, and [3]Paediatric Dentistry, Barts and The London Dental School, Queen Mary, University of London, London E1 2AD, UK

*International Journal of Paediatric Dentistry 2009; 19: 219–221*

## Introduction

Lingual glial choristoma is a rare developmental lesion. A choristoma is a mass of histologically normal tissue present in an abnormal anatomical location. Glial cells are normally found within the central nervous system, and provide an insulatory and supportive role for the neuronal cells.

This kind of choristoma is very rare having been reported in the literature on only 14 previous occasions. It was first reported by Peterer in 1922[1] and subsequently by other authors. Presentation with this lesion as the literature shows is often in the first few days or weeks of life. The main presenting complaints are airway obstruction or feeding difficulties due to the size of the lesion. Management of the lesion is usually by surgical excision.

## Case report

A 3-year-old girl of mixed race (white Caucasian and Afro-caribbean) was referred to The Royal London Hospital regarding an asymptomatic swelling on her tongue.

Clinical examination revealed a 5 mm × 5 mm × 5 mm, pedunculated mucosal lesion on the dorsal surface of the tongue, to the left of the midline, and anterior to the junction between posterior one-third and anterior two-thirds of the tongue. The lesion was firm on palpation and covered with an intact non-ulcerated mucosa. There was no other mucosal pathology (Fig. 1).

Under general anaesthesia, an excision biopsy of the lesion was performed using mono-polar electrocautery.

A haematoxylin-and-eosin-stained section of the biopsy revealed a nodular piece of lingual mucosa with variably atrophic lingual papillae. The corium showed an ill-defined accumulation of faintly myxoid tissue with scattered spindle-shaped and stellate cells (Fig. 2). At higher magnification, the background myxoid tissue appeared to consist of a tangled meshwork of fibrillary processes of the stellate and spindle-shaped cells. Focally dystrophic calcification was seen.

Immunohistochemistry revealed strong expression of glial fibrillary acidic protein (GFAP) in the lesional cells (Fig. 3). Heterogeneous expression of S100 protein was seen. GFAP highlighted the tangled cell processes of the stellate and bipolar cells. Expression of neurofilament

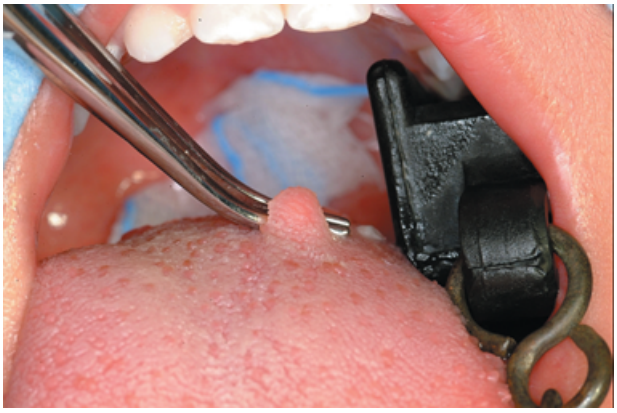

**Fig. 1.** Pre-operative appearance of the lump on the dorsal surface of the tongue.

*Correspondence to:*
David James Baldwin, Paediatric Dentistry, c/o Room 211, 2nd Floor, Dental Institute, The Royal London Hospital, New Road, Whitechapel, London E1 1BB, UK. E-mail: david.baldwin@bartsandthelondon.nhs.uk

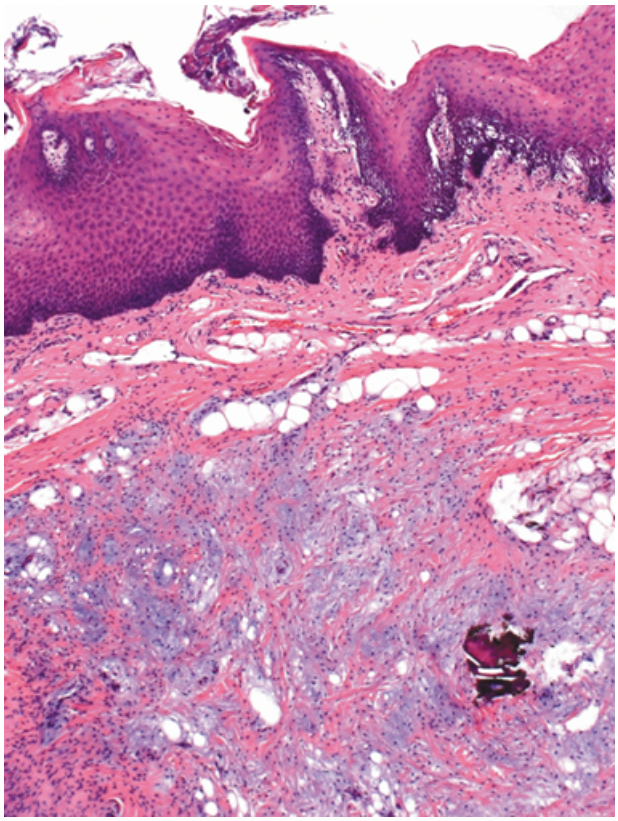

**Fig. 2.** Lingual mucosa showing an ill-defined area of faintly myxoid appearing tissue with scattered spindle-shaped cells, interspersed with clusters of adipocytes. Focally dystrophic calcification is seen. (Haematoxylin and eosin ×10).

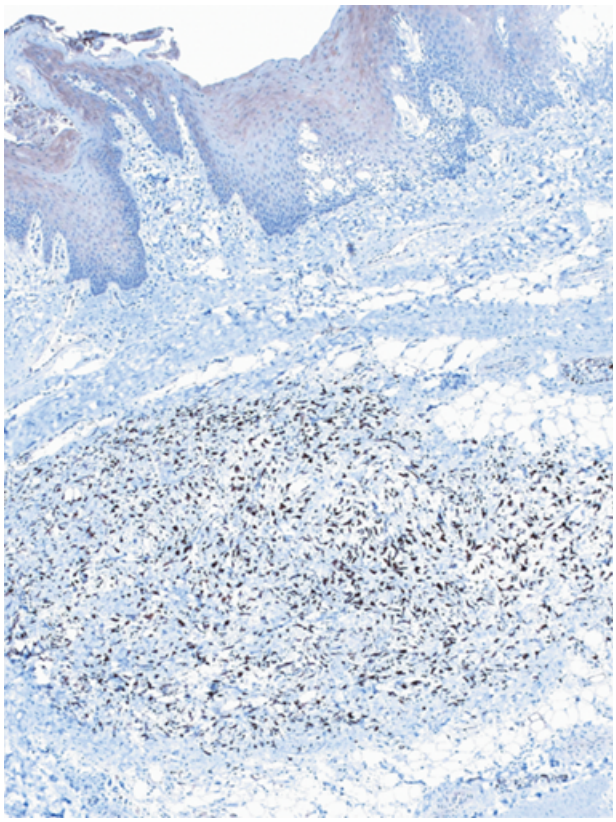

**Fig. 3.** Immuno-histochemically, the spindle-shaped cells express glial fibrillary acidic protein. (Glial fibrillary acidic protein ×10).

(NF) protein was seen only in the nerve fibre bundles of the surrounding tissue. The diagnosis rendered was of heterotopic glial tissue (glial choristoma).

On review, at 2 and 6 months, the excision site had healed well with no complication or recurrence of the lesion.

## *Comment*

The reason for excision was to attain a definitive diagnosis in order to ease the patient's parents' concerns over the cause of the lesion. A number of differential diagnoses were considered: mucosal tag due to trauma, fibro-epithelial polyp, viral papilloma, neurofibroma, and vascular lesion, but none of these were considered definitively to be the cause of the lesion.

Excision was carried out using electrocautery to remove the lesion, obtain haemostasis, and avoid needing sutures. Alternatively, excision with cold steel followed by electrocautery to achieve haemostasis could have been used. The latter technique would have allowed histological examination of the margins to show whether complete excision had been achieved (Table 1).

When diagnosis of a lesion is uncertain, the authors would strongly recommend scalpel excision in order to allow histological examination of the resection margins and to enable the pathologist to confirm complete removal of the lesion.

Most reported cases of heterotopic brain tissue consist of glial tissue with only a small proportion containing neurons[2,3]. Astrocytes comprise the predominant and often the only component in most cases. Other structures that may be seen are ependyma-lined clefts, choroid plexus, and retinal structures, usually in the palato-pharyngeal cases[4]. In our case, stellate and bipolar astrocytes were the only cell type seen.

**Table 1. A comparison of surgical treatment options.**

| | Advantages | Disadvantages |
|---|---|---|
| Cold steel | Quick and simple to use | Causes bleeding |
| | Good proprioceptive feedback for surgeon | Needs sutures to close excision site |
| | Non-obliterated margins | |
| Electrocautery | Quick and simple to use | Poor proprioceptive feedback for surgeon |
| | Coagulation with excision | Obliterated margins of excision – confirmation of removal of whole lesion not possible |
| | No need to suture | |

Neoplastic transformation is not seen in lingual cases, but has been reported rarely in palatopharyngeal cases[4].

In most cases, histological diagnosis is straightforward on a haematoxylin-and-eosin-stained section, if one is aware of the entity.

Immunohistochemically, antibodies to GFAP demonstrate the intermediate filament proteins of the astrocytes. Certain studies have reported expression of CD57, weak, focal NF and neuron-specific enolase as well as proliferating cell nuclear antigen in one case[5]. In our case, the histological appearance together with the clinical features was diagnostic of a glial heterotopia. Staining for antibodies to GFAP demonstrated the cytologic features with the multipolar and bipolar processes.

Pathogenesis of glial choristoma is speculated to be due to separation of pluripotential embryonic cells from the developing central nervous system before complete fusion of the neural tube and their migration with cranial nerves and muscle into the oral cavity[2,3].

According to some reports[2,6], glial choristomas can grow, sometimes rapidly; early diagnosis in this case therefore allowed very simple surgical excision with little discomfort to the patient. Larger lesions have on occasion caused concern when affecting the breathing and feeding of a patient thus prompting removal. The prognosis for the patient, however, is excellent as these are developmental, non-neoplastic lesions and do not usually recur following surgical excision.

**What this paper adds**

- Documentation of a rare developmental lesion affecting the oral cavity
- To add to the current data in the literature on lingual glial choristoma
- Validates surgical excision as the optimum treatment modality for this type of lesion
- To discuss histopathological techniques to confirm diagnosis of glial choristoma

**Why this paper is important to paediatric dentists**

- Highlights glial choristoma as a soft tissue lesion that can present in the mouth
- Raises awareness of an alternative differential diagnosis for lingual pathology
- Addresses various management options for soft tissue lesions in infants
- Illustrates the importance of close liaison with other specialities in the management of the patient – in this case, the oral pathologist

## References

1 Peterer F. Uber glioma linguae. *Frankf Z Pathol* 1922; **26**: 214–226.
2 Abdelsayed RA, Wetherigton W, Bent JP, Sharpe DE. Glial choristoma of the tongue. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod* 1999; **87**: 215–222.
3 Takamizawa S, Inoue T, Ono Y, *et al*. A case report of glial choristoma of the tongue. *J Pediatr Surg* 2006; **41**: E13–E15.
4 Rosenblum MK, Bilbao JM, Ang LC. Neuromuscular system. In: Rosai J (ed.). *Ackerman's Surgical Pathology*, 9th edn, Vol. 2. London: Mosby, 2004: 2465.
5 Ide F, Shimoyama T, Horie N. Glial choristoma in the oral cavity: histopathologic and immunohistochemical features. *J Oral Pathol Med* 1997; **26**: 147–150.
6 Odell EW, Morgan PR. *Biopsy Pathology of the Oral Tissues*, 1st edn. London: Chapman & Hall, 1998: 141–142.