CASE REPORT

Rare glomangiosarcoma of the tongue

Sumetha Rajendran,¹ Arthur Harry Henderson,² Stuart Gillett²

¹Oral and Maxillofacial Surgery, University of Bristol Medical School, Bristol, UK
²Oral and Maxillofacial Surgery, Royal United Hospital Bath NHS Trust, Bath, UK

Correspondence to
 Dr Sumetha Rajendran,
 sumetha.rajendran@hotmail.com

Accepted 23 June 2018

SUMMARY

The glomus tumour is a rare neoplasm derived from the glomus apparatus. Subungual sites are most common with only three published cases involving the dorsal tongue. To our knowledge, this is the first case of an intraoral malignant glomus tumour (glomangiosarcoma) to be published in English literature. We report a case of a single glomus tumour located on the posterior dorsal tongue of a middle-aged man, which was surgically excised. Immunohistological features were indicative of a glomus tumour; however, in this case, malignant features were also discovered, warranting re-excision.

BACKGROUND

The glomus tumour is a usually benign tumour, which arises from the glomus apparatus, an arteriovenous anastomosis situated in the dermis. The purpose of the glomus apparatus is to regulate blood flow via the anastomosis and contribute to thermal regulation.¹ It is most commonly located in the fingers and nail beds, where the majority of glomus tumours arise.² The occurrence of a glomus tumour in the oral cavity is very rare, and presentation is usually painless with no sex predilection. The aetiology of this neoplasm is unknown, though familial glomus tumours have been known to follow an autosomal dominant pattern.

CASE PRESENTATION

A 51-year-old Caucasian man presented to the oral and maxillofacial surgery department with a 2-month history of right-sided posterior tongue discomfort. There was no associated pain, bleeding or ulceration and no reported dysphagia. He was a non-smoker with no relevant medical or family history. On examination, a firm, discrete submucosal mass was palpable within the substance of the right dorsal surface of the tongue. The surrounding mucosa displayed no pathology, and there was no palpable cervical lymphadenopathy.

INVESTIGATIONS

The lesion was investigated with MRI at first instance, which showed a well-defined ovoid mass within the right hemitongue involving the intrinsic musculature, with no evidence of disease elsewhere (figure 1). Fine-needle aspiration cytology (FNA) showed some dark staining groups of cohesive epithelioid cells, and the patient was referred to the ENT department for a diagnostic biopsy.

Biopsy may be more useful for a clear diagnosis but carries a higher risk of bleeding in such a highly vascular lesion. FNA in a vascular lesion such as this

carries a lower risk of bleeding but also introduces diagnostic uncertainty or the possibility of a non-diagnostic sample.

H&E staining of the biopsy specimen revealed a vascular lesion containing medium-sized cells in sheet-like growth pattern with mildly pleomorphic nuclei, closely related to the blood vessels within the specimen. The cells showed strong positivity with vimentin and smooth muscle actin, and with desmin in a proportion of cells. Overall, the result was consistent with a glomus tumour.

DIFFERENTIAL DIAGNOSIS

On visual inspection, this tumour resembles a vascular lesion such as a haemangioma or haemangiopericytoma. The clinical features of this tumour overlap with pericyte derivatives, such as cavernous haemangioma, epithelioid haemangioma and myopericytoma³; however, histologically, these differentials can be excluded due to the lack of glomus cells. Prior to Masson (1924), this tumour was thought to be an angiosarcoma variant. However, on histological analysis, Masson revealed the ultrastructure of this tumour resembles that of the modified smooth muscle cells of the normal glomus body and comes under the category of a smooth muscle tumour.⁴

TREATMENT

The lesion was uneventfully excised, and the 27×24×14 mm gross specimen was analysed extensively due to atypical features (figure 2). Light microscopic evaluation of H&E-stained sections showed tumour with a fibrous capsule and sheets of small cells that contained round to ovoid mildly pleomorphic nuclei and abundant cytoplasm. Mitotic figures (3 per 10 high-power fields) were identified and several foci of lymphovascular space invasion were present. Some foci of tumour cells had pushing and infiltrative edges, and in some places abutted the inked surgical margins. The cells were arranged with slit-like vessels within the lesion, and a prominence in the background vascularity could be seen with focal staghorn blood vessels present.

Immunostaining once again showed strong positivity with smooth muscle actin and vimentin and a variable degree of positivity with desmin. H-Caldesmon was positive in several foci and CD34 showed weak and focal positivity within the tumour cells. There was negative staining for CD31, AE₁, AE₂ and MNF116. The proliferation fraction as assessed by Ki-67 staining was approximately 15%–20%.

The immunohistochemical profile showed features consistent with a glomus tumour diagnosis;

© BMJ Publishing Group Limited. No commercial re-use. See rights and permissions. Published by BMJ.

To cite: Rajendran S, Henderson AH, Gillett S. *BMJ Case Rep* Published Online First: [please include Day Month Year]. doi:10.1136/bcr-2017-223268

Figure 1 MRI head and neck showing well-defined 23.65 mm × 13.27 mm lesion in right hemitongue of increased signal on T2 and short inversion time inversion recovery (STIR) imaging. Intrinsic and extrinsic tongue muscles involved, however, do not quite reach the midline or surface of the tongue. No pathological lymphadenopathy, no salivary gland involvement.

however, it also highlighted concerning features. This appeared to be a highly cellular tumour demonstrating vascular invasion and increased mitotic figures (figure 3). The morphological features were indicative of the malignant variant of the glomus tumour, the glomangiosarcoma. Further, it could not be conclusively reported as fully excised due to the positive inked surgical margins. This, added to the malignant features, prompted discussion as to whether further surgery would be advisable (figure 4).

OUTCOME AND FOLLOW-UP

Following analysis of the histopathology reports and an multi-disciplinary team (MDT) meeting, the patient was offered wider excision. He declined surgery initially preferring regular follow-up; however, some months later he consented to excision of the tumour margins. Following successful scar excision with no evidence of residual tumour, no future surgical intervention was indicated.

DISCUSSION

Glomus tumours are usually benign soft-tissue tumours, most commonly found in the subungual regions of the fingers and toes, the palm of the hand, the wrist and the forearm. There are also reported cases involving the trachea, lung, heart, stomach

and uterus.⁵ Intraoral cases are extremely rare, due to the very nature of the tumour. The glomus body lies within the stratum reticularis of the dermis and is therefore most frequently found in subungual areas. It was studied in 1924 by Masson, who was the first to publish findings that contradicted the view that the glomus tumour was a form of angiosarcoma. He observed that the tumour, in comparison with the normal glomus body, potentially represented abnormal growth of the structure. The normal glomus apparatus consists of a layer of glomus cells surrounding specialised blood vessels. These form the Sucquet-Hoyer canal, which contains the arteriovenous anastomosis.⁶ It was shown by Masson that the origin of the neoplastic process that results in the glomus tumour was the epithelioid glomus cell surrounding the Sucquet-Hoyer canal.

Usually, the tumour appears as a small, slow-growing, painless mass that can be treated with local excision; however, Spector *et al* reported malignant glomus tumours in the head and neck region exhibiting intracranial spread.⁷ A case of glomangiosarcoma metastasising from a distant site to the jaw was published in 1987 by Kreutz *et al*.⁸ He reported a case of a glomangiosarcoma

Figure 2 Intraoperative image of glomus tumour excision.

Figure 3 Tumour histology showing a highly cellular tumour containing sheets of small cells with ovoid nuclei that are mildly pleomorphic. Mitotic figures of up to 3 per 10 high-power fields are present. Lesional cells are set within a well-vascularised fibrous stroma.

Figure 4 Postoperative image of fully excised glomus tumour, margins needed re-excision following histology report.

of the lateral thigh metastasising to the maxillary tuberosity. This was even more unusual due to the rarity of metastases in this area: 1% of malignant neoplasms metastasise to the jaw and only 1% of oral malignancies are metastases. Specific figures for the prevalence of metastasis for glomangiosarcoma are still unknown due to the rarity of this entity and lack of published data. To the best of the authors' knowledge, this is the first published case of a malignant intraoral glomus tumour.

The glomangiosarcoma accounts for only 1% of all glomus tumours and is therefore an incredibly rare malignant variant. Folpe *et al* in 2001 proposed classification criteria for glomangiosarcomas, based on the histology, which included a size of more than 2 cm and a deep location, or atypical mitotic figures, or more than five mitotic figures per 50 high-power fields⁹; this case fulfils his criteria. The classic glomus tumour has two benign variants, the glomangioma and glomangiomyoma, with the solid glomus tumour presenting most commonly.¹⁰ Glomangiomas have a predominantly vascular component with relatively few glomus cells present within the walls. Glomangiomyomas predominantly consist of smooth muscle, which is prominent either in combination with a glomus tumour pattern or glomangioma.

The solid glomus tumour tends to demonstrate a capsule and ultrastructurally shows features of smooth muscle, in this case immunostaining highlighted strong positivity with smooth muscle actin. It has been debated as to whether the origin of the glomus tumour is smooth muscle or pericytes. Glomus cells have been demonstrated to have ultrastructural features of both¹¹; however, most analyses have concluded that glomus cell characteristics most resemble those of smooth muscle cells. Rallis *et al* stated that positive staining for smooth muscle actin and vimentin, and negative staining for S-100 and epithelial markers are necessary to verify the diagnosis.¹² The tumour cells were positive for vimentin and desmin intermediate filaments consistent with smooth muscle cells; however, previous studies show mostly negative staining for desmin in glomus cells.^{13 14} An explanation for the patchy positivity for desmin seen here and in other cases could be differentiation to vascular smooth muscle cells, as desmin is also absent in aortic smooth muscle cells.¹⁵

The tumour was negative for factor XIII, CD31 and CD34. The absence of endothelial cell marker factor VIII is consistent with previous studies of glomus cells. Moody *et al* presented a series of 20 glomus tumours, all of which were factor VIII negative, with factor VIII antigen consistently related to the endothelial cells only.¹⁶ Absence of endothelial markers CD31

and CD34 rule out a tumour of vascular origin. Concerning features of raised mitotic figures, lymphovascular space invasion and increased proliferation fraction highlighted the possibility of malignancy. It has been argued that due to the lack of evidence for metastasis of any glomus tumour in the oral cavity, the malignant variety may not exist in the true definition.⁵ Despite the lack of published evidence with regards to metastatic spread, this possibility should not be overlooked. It is therefore important to perform immunohistochemical analysis taking note of atypical features, though this tumour rarely acts aggressively.

Conclusion

In summary, we have presented the case of a slow-growing, non-tender lump in the posterior dorsal tongue of a middle-aged man. Biopsy results highlighted a well-circumscribed, highly vascular tumour with characteristic glomus cells in sheet-like formation. However, detailed analysis identified malignant features such as raised mitotic index, increased proliferation rates and lymphovascular space invasion, resulting in glomangiosarcoma being the most likely diagnosis, and warranting wider excision. This very unusual case aids our understanding of this rare clinical entity and highlights the benefit of an MDT approach to facilitate the best treatment plan for this patient. Intraoral glomus tumours have consistently been described as benign tumours, with glomangiosarcoma being an incredibly rare variant; however, this case highlights the possibility of metastatic spread and the importance of identifying malignant features.

Learning points

- Glomus tumours are rare neoplasms that are mostly benign, slow growing and painless in the oral cavity.
- They are usually managed with local excision, and rarely recur or metastasise.
- Glomangiosarcomas are incredibly rare with this being the first published intraoral case in English literature.
- Identifying malignant features on immunohistochemical analysis allows a decision to be made with regards to indications for re-excision and risk of metastasis.

Contributors SR: case report write-up. AHH: clinical photos, patient consent form and final draft edit. SG: own surgical case, provided outpatient notes and photos.

Funding The authors have not declared a specific grant for this research from any funding agency in the public, commercial or not-for-profit sectors.

Competing interests None declared.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

REFERENCES

- 1 Kaplan IW, Karlin S. Glomus tumor; a report of two cases. *Am J Surg* 1953;86:192.
- 2 Calonje E, Fletcher CD. Cutaneous intraneural glomus tumor. *Am J Dermatopathol* 1995;17:395–8.
- 3 Kessaris P, Klimis T, Zanakos S. Glomus tumour of the hard palate: case report and review. *Br J Oral Maxillofac Surg* 2001;39:478–9.
- 4 Weiss SW, Goldblum JR, Enzinger and Weiss's soft tissue tumours, 2001:985–96.
- 5 Enzinger FM, Weiss SW. *Soft tissue tumours*. St. Louis, MO: Mosby, 1983:450–60.
- 6 Masson P. Le glomus neuromyoarterial des regions tactiles et ses tumeurs. *Lyon Chir* 1924;21:257.
- 7 Spector GJ, Ciralsky RH, Ogura JH. Glomus tumors in the head and neck: III. Analysis of clinical manifestations. *Ann Otol Rhinol Laryngol* 1975;84:73–9.
- 8 Kreutz RW, Christensen RE, Fish LR. Glomangiosarcoma with metastasis to the maxilla. *Int J Oral Maxillofac Surg* 1987;16:116–8.
- 9 Folpe AL, Fanburg-Smith JC, Miettinen M, *et al*. Atypical and malignant glomus tumors: analysis of 52 cases, with a proposal for the reclassification of glomus tumors. *Am J Surg Pathol* 2001;25:1–12.

- 10 Fletcher DC, Unni KK, Mertens F. *Glomus tumours, in World Health Organisation Classification of Tumours, Pathology and Genetics of Tumours of Soft Tissue and Bone*. Lyon: IARC Press, 2002:137.
- 11 Osamura RY, Watanabe K, Yoneyama K, *et al*. Glomus tumor of the stomach. Light and electron microscopic study with literature review of related tumors. *Acta Pathol Jpn* 1977;27:533–9.
- 12 Rallis G, Komis C, Mahera H. Glomus tumor: a rare location in the upper lip. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod* 2004;98:327–36.
- 13 Dervan PA, Tobbia IN, Casey M, *et al*. Glomus tumours: an immunohistochemical profile of 11 cases. *Histopathology* 1989;14:483–91.
- 14 Miettinen M, Paal E, Lasota J, *et al*. Gastrointestinal glomus tumors: a clinicopathologic, immunohistochemical, and molecular genetic study of 32 cases. *Am J Surg Pathol* 2002;26:301–11.
- 15 Frank ED, Warren L. Aortic smooth muscle cells contain vimentin instead of desmin. *Proc Natl Acad Sci U S A* 1981;78:3020–4.
- 16 Moody GH, Myskow M, Musgrove C. Glomus tumor of the lip. A case report and immunohistochemical study. *Oral Surg Oral Med Oral Pathol* 1986;62:312–8.

Copyright 2018 BMJ Publishing Group. All rights reserved. For permission to reuse any of this content visit <http://group.bmj.com/group/rights-licensing/permissions>.
BMJ Case Report Fellows may re-use this article for personal use and teaching without any further permission.

Become a Fellow of BMJ Case Reports today and you can:

- Submit as many cases as you like
- Enjoy fast sympathetic peer review and rapid publication of accepted articles
- Access all the published articles
- Re-use any of the published material for personal use and teaching without further permission

For information on Institutional Fellowships contact consortiasales@bmjgroup.com

Visit casereports.bmj.com for more articles like this and to become a Fellow