A Rare Case of Glomus Tumor on the Mucosal Surface of Lower Lip

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Abstract
Glomus tumors are mesenchymal neoplasms derived from glomus bodies with rare presentations in the oral cavity. Glomus tumors present as a purple or pink vascular nodule or papule, sized < 1 cm, and imitate vascular neoplasms such as hemangiopericytoma or hemangioma. Glomus tumors represent less than 2% of all benign soft tissue tumors. Only 27 cases of benign glomus tumors with oral cavity involvement have been reported to date. The most-reported oral tumors involved the lips (54.2%), followed by hard palate, gingiva, tongue, and buccal mucosa. The mean age of presentation of the labial glomus tumors is 48.7 years, with no gender predilection, in contrast to the subungual site, which occurs more in females. The etiology of the glomus tumors remains unknown. Subungual glomus tumors present as stabbing pain, cold intolerance, and tenderness of the fingertips, whereas labial glomus tumors mostly present as a painless, small, and slow-growing lesion. Treatment is surgical resection of the tumor. The recurrence rate of labial glomus tumors is unclear. In this article, we present the case of a 62-year-old man with a 2-month history of painless, soft lump on the mucosal surface of the lower left lip. Excisional resection of the tumor was performed in the clinic, and the histopathologic finding was consistent with solid glomus tumor. At 1 year follow-up there was no recurrence.

Keywords
glomus tumor, oral cavity, labial glomus tumor

Introduction
Glomus tumors are mesenchymal neoplasms derived from glomus bodies. The glomus bodies are located between the venous and arterial systems in the subungual dermis of the digits. The role of glomus bodies is thermoregulation in response to temperature changes. Glomus tumors present as a purple or pink vascular nodule or papule that can imitate a vascular neoplasm such as hemangiopericytoma or hemangioma. Initially, glomus tumors were considered as a variant of angiosarcoma. In 1924, Masson published his findings that revealed that glomus tumors are histologically similar to smooth muscle cells of the normal glomus bodies. Since then glomus tumors are considered under the category of smooth muscle tumors. The occurrence of glomus tumors in the oral cavity is rare. In this article, we present a case with a 2-month history of a 1 cm lesion on the lower-left labial mucosa of the lip.

Case Presentation
A 62-year-old man presented to the dental clinic with a 2-month history of painless, round, and nonerosive lump on the inner surface of the lower left lip. The patient reported that he first noticed a small lump on the lip, which gradually increased in size up to 1 cm. He denied any history of trauma, ulceration, drainage, and bleeding. On examination, a 1-cm round, nontender, and mobile lump on the lower left labial mucosa was observed. The remainder of the physical examination and review of systems were unremarkable. His past medical history included diabetes mellitus type 2. Family history was unremarkable for any type of malignancy in his first-degree relatives. Medication history included metformin and aspirin. He was a former smoker, half a pack per day for 12 years. He quit smoking 6 years ago when he was diagnosed with diabetes. He denied alcohol and drug use. The patient consented for an excisional biopsy. The lesion was excised under local anesthesia in the clinic. Histopathologic examination revealed submucosal proliferation of monotonous, bland compact epithelioid cells arranged in sheets, and punctuated by blood vessels suggesting glomus tumors.
There was no atypia or malignant cells (Figures 1A and 2). Immunostaining with smooth muscle actin was diffusely positive, which supported the diagnosis of solid glomus tumors (Figure 1B). The patient followed up with us after 1 year and there was no recurrence of the tumor.

**Discussion**

Glomus tumors are rare mesenchymal neoplasm, which usually present in subungual areas of the toes and fingers. Glomus tumors represent <2% of all benign soft tissue tumors. Extraluminal presentations of glomus tumors are very rare; less than 1% of glomus tumors have been found on the head region. The most-reported sites of oral glomus tumors are lips (54.2%), followed by hard palate, gingiva, tongue, and buccal mucosa. Few cases have been reported with the involvement of the lung, trachea, heart, uterus, and stomach.

Glomus tumors are subcategorized as solid (75% of cases), glomangiomas, with vascular predominance (20%) and glomangiomyomas, with smooth muscle cell predominance (5%), depending on the proportion of glomus cells, blood vessels, and smooth muscle.

The size of glomus tumors in the dermis or subcutaneous tissues of extremities is usually <1 cm. However, the size of glomus tumors in the head and neck area are larger and averaged 1 to 1.5 cm. There are no data suggesting that tumor size influences the patients’ prognosis.

Few cases of malignant glomus tumors have been reported. A malignant variant of glomus tumors is glomangiosarcoma. Malignancies in the head and neck area are very rare. Spector et al reported a case with glomangiosarcoma in the head and neck with intracranial metastasis. Kreutz et al published a case with glomangiosarcoma metastasis to the jaw from a distant location. The diagnosis of malignant glomus tumors is based on histopathological examination. Tumors with a size of more than 2 cm, moderate-to-high nuclear grade, and atypical mitotic figure more than 5 per 50 HPF (high-power field) will be considered highly suspicious for malignancy.

Folpe et al in 2001 after analysis of 52 cases of glomus tumors and combining the histological features proposed...
classification of glomus tumors with atypical features: malignant glomus tumors (large size and deep location, marked atypia with mitotic activity, as glomangiosarcoma), symplastic glomus tumor (only nuclear atypia), glomus tumor of uncertain malignant potential (superficial location with high-mitotic activity or large size only or deep location only), and glomangiomatosis (histological features of benign glomus tumor with diffuse growth).7

The etiology of the glomus tumors remains unclear; however, familial glomus tumors found to have an autosomal dominant pattern with incomplete penetration.13 Yoo et al14 reported a case of double glomus tumors in the submandibular and parotid regions. Sixty percent of patients with multiple glomus tumors have had a positive family history, supporting the concept that this abnormality is an inherited disease.15 The first case of the glomus tumor of the oral cavity was reported in 1943. To the best of our knowledge, only 27 cases of benign oral glomus tumors have been reported until now, in a review of the English-language literature (Table 1). Rajendran et al19 reported a first intraoral case of glomangiosarcoma in English literature: a 51-year-old male with a glomangiosarcoma of tongue in size of 27 mm.

Subungual glomus tumors present as stabbing pain, cold intolerance, and tenderness of the fingertips, whereas labial glomus tumors mostly present as a painless, small, slow-growing lesion.12,40 Glomus tumors usually appear in patients aged 40 to 70 years old.41 Data show that the average age of presentation of the labial tumors is 48.7 years without sex predilection in contrast to the subungual site, which occurs more in females. However, other regions have shown even distribution between sexes.10,42 Treatment is surgical resection of the tumor. The recurrence rate of subungual tumors is 4% to 15%, whereas there are insufficient documents to estimate the recurrence rate of labial glomus tumors.43,44

**Conclusion**

Labial glomus tumors have different clinical presentations compared to subungual glomus tumors. This difference in clinical presentation makes it difficult for clinicians to differentiate these tumors from other more common painless lesions of the lip. Glomus tumors should be in the differential diagnosis for well-circumscribed, nontender lesions in the oral cavity. We hope this article would help clinicians to be more aware of this rare disease.

**Table 1. Characteristics of cases with benign oral glomus tumors.**

| Case | Year | Author | Age (years) | Gender | Location | Symptoms | Size (mm) |
|------|------|--------|-------------|--------|----------|----------|-----------|
| 1    | 1954 | King16 | 32          | Male   | Gingiva  | Tenderness | 6         |
| 2    | 1965 | Harris and Griffin17 | 35 | Female | Periodontium | Pain | 5 |
| 3    | 1967 | Sidhu18 | 10 | Female | Hard palate | Unknown | Unknown |
| 4    | 1976 | Charles19 | 17 | Female | Hard palate | No | Unknown |
| 5    | 1979 | Sato et al20 | 29 | Male | Tongue | No | 3 |
| 6    | 1981 | Tajima et al21 | 63 | Female | Tongue | No | Unknown |
| 7    | 1985 | Saku et al22 | 45 | Male | Buccal mucosa | No | 45 |
| 8    | 1986 | Ficarra et al23 | 51 | Female | Upper lip (mucosa) | No | 20 |
| 9    | 1986 | Moody et al24 | 65 | Female | Upper lip | No | 10 |
| 10   | 1987 | Staicic and Bojic25 | 55 | Male | Tongue | Unknown | Unknown |
| 11   | 1992 | Geraghty et al26 | 71 | Male | Hard palate | No | 15 |
| 12   | 1995 | Kusama et al27 | 57 | M | Upper lip (mucosa) | Tenderness | Unknown |
| 13   | 1996 | Savaci et al28 | 55 | Female | Buccal mucosa | Pain | 10 |
| 14   | 1997 | Sakashita et al29 | 54 | Male | Upper lip (mucosa) | No | 12 |
| 15   | 2000 | Yu et al30 | 54 | Female | Face, lower lip, buccal mucosa | No | Unknown |
| 16   | 2001 | Kessaris et al3 | 46 | Female | Hard palate | No | 18 |
| 17   | 2004 | Rallis et al31 | 85 | Female | Upper lip (mucosa) | Pain | 13 |
| 18   | 2005 | Lanza et al32 | 65 | Male | Lower lip | Unknown | Unknown |
| 19   | 2008 | Ide et al33 | 57 | Male | Upper lip | Unknown | 8 |
| 20   | 2008 | Ide et al33 | 54 | Male | Upper lip | Unknown | 12 |
| 21   | 2010 | Boros et al10 | 34 | Male | Lower lip (mucosa) | No | 15 |
| 22   | 2010 | Dérand et al34 | 11 | Female | Lower lip (vermilion) | No | 3 |
| 23   | 2018 | Vasconcelos et al35 | 67 | Female | Upper lip (mucosa) | Pain | 10 |
| 24   | 2018 | Sánchez-Romero et al36 | 51 | Female | Upper lip (mucosa) | Pain | 10 |
| 25   | 2018 | Smith et al37 | 26 | Male | Lower lip | Pain | 15 |
| 26   | 2018 | Smith et al37 | 58 | Female | Tongue | No | 20 |
| 27   | 2018 | Zou et al38 | 24 | Male | The floor of the mouth | Pain | 28 |
| 28   | 2019 | Current case | 62 | Male | Lower Lip (mucosa) | No | 10 |
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Ethics Approval
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Informed Consent
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References
1. Naik SM, Shenoy AM, Nanjundappa, et al. Paragangliomas of the carotid body: current management protocols and review of literature. Indian J Surg Oncol. 2013;4:305-312.
2. Gombos Z, Zhang PJ. Glomus tumor. Arch Pathol Lab Med. 2008;132:1448-1452.
3. Keswasri P, Klimis T, Zanakis S. Glomus tumour of the hard palate: case report and review. Br J Oral Maxillofac Surg. 2001;39:478-479.
4. Weiss SW, Goldblum JR. Enzinger and Weiss's Soft Tissue Tumors. 6th ed. Saunders; 2001:985-996.
5. Shugart RR, Soule EH, Johnson EW Jr. Glomus tumor. Surg Gynecol Obstet. 1963;117:334-340.
6. Makino Y. A clinicopathological study on soft tissue tumors of the head and neck. Acta Pathol Jpn. 1979;29:389-408.
7. Folpe AL, Fanburgsmith JC, Miettinen M, Weiss SW. 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.
8. Enzinger FM, Weiss SW. Soft Tissue Tumors. 6th ed. Saunders; 2014:749-759.
9. Pulitzer DR, Martin PC, Reed RJ. Epithelioid glomus tumor. Hum Pathol. 1995;26:1022-1027.
10. Boros AL, Davis JP, Sedghizadeh PP, Yamashita DDR. Glomus tumor: report of a rare case affecting the oral cavity and review of the literature. J Oral Maxillofac Surg. 2010;68:2329-2334.
11. Spector GJ, Ciralsky RH, Ogru JA. Glomus tumors in the head and neck: III. Analysis of clinical manifestations. Ann Otol Rhinol Laryngol. 1975;84:73-79.
12. Kreutz RW, Christensen RE, Fish LR. Glomangiosarcoma with metastasis to the maxilla. Int J Oral Maxillofac Surg. 1987;16:116-118.
13. Troschke A, Weyers W, Schill WB. Multiple familial glomangioma [in German]. Hautarzt. 1993;44:731-734.
14. Yoo YS, Choi JH, Heo G, Kim SW, Kim HJ. Double glomus tumors originating in the submandibular and parotid regions. Clin Exp Otorhinolaryngol. 2011;4:49-51.
15. Boon LM, Brouillard P, Irtithun A, et al. A gene for inherited cutaneous venous anomalies (“glomangiomas”) localizes to chromosome 1p21-22. Am J Hum Genet. 1999;65:125-133.
16. King ES. Glomus tumor. Aust N Z J Surg. 1954;23:280-295.
17. Harris R, Griffin CJ. Glomus tumor of the periodontal tissues. Aust Dent J. 1965;10:33-37.
18. Sidhu SS. Glomus tumor of palate. Indian Dent Assoc. 1967;39:167-168.
19. Charles NC. Multiple glomus tumors of the face and eyelid. Arch Ophthalmol. 1976;94:1283-1285.
20. Sato M, Shirasuna K, Sakuda M, et al. Fine structure of a glomus tumor of the tongue and expression of C type virus in its tumor cells. Int J Oral Surg. 1979;8:199-204.
21. Tajima Y, Weathers DR, Neville BW, Benoit PW, Pedley DM. Glomus tumor (glomangioma) of the tongue: a light and electron microscopic study. Oral Surg Oral Med Oral Pathol. 1981;52:288-293.
22. Saku T, Okabe H, Matsutani K, Sasaki M. Glomus tumor of the cheek: an immunohistochemical demonstration of actin and myosin. Oral Surg Oral Med Oral Pathol. 1985;60:65-71.
23. Ficarra G, Merrell PW, Johnston WH, Hansen LS. Intraoral solitary glomus tumor (glomangioma): case report and literature review. Oral Surg Oral Med Oral Pathol. 1986;62:306-311.
24. Moody GH, Myskow M, Musgrove C. Glomus tumor of the lip. A case report and immunohistochemical study. Oral Surg Oral Med Oral Pathol. 1986;62:312-318.
25. Stajicic Z, Bojic P. Intraoral glomus tumour. A case report. J Craniomaxillofac Surg. 1987;15:376-378.
26. Geraghty JM, Thomas RW, Robertson JM, Blundell JW. Glomus tumour of the palate: case report and review of the literature. Br J Oral Maxillofac Surg. 1992;30:398-400.
27. Kusama K, Chu L, Kidokoro Y, et al. Glomus tumor of the upper lip. J Nihon Univ Sch Dent. 1995;37:97-101.
28. Savaci N, Emiroglu M, Gunmen M, Gungor S. A rare case of glomus tumor; buccal localization. Br J Oral Maxillofac Surg. 1996;34:199-200.
29. Sakashita H, Miyata M, Nagao K. Glomus tumor in the upper lip. A case report. Int J Oral Maxillofac Surg. 1997;26:301-302.
30. Yu HH, Kwon SJ, Bahn JY, Park JM, Park YM. Localized multiple glomus tumors of the face and oral mucosa. J Dermatol. 2000;27:211-213.
31. 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;98:327-336.
32. Lanza A, Moscariello A, Villani R, Colella G. Glomus tumor of the lower lip. A case report [in Italian]. Minerva Stomatol. 2005;54:678-690.
33. Ide F, Mishima K, Yamada H, et al. Periocular myoid tumors of the orbital region: a clinicopathologic re-evaluation of 35 cases. J Oral Pathol Med. 2008;37:43-49.
34. Dérand P, Warfvinge G, Thor A. Glomangioma: a case presentation. J Oral Maxillofac Surg. 2010;68:204-207.
35. Vasconcelos ACU, Loyola AM, Gomes APN, et al. A symptomatic swelling of the upper lip. Oral Surg Oral Med Oral Pathol Oral Radiol. 2018;125:107-111.
36. Sánchez-Romero C, Oliveira MEP, Castro JFL. Glomus tumor of the oral cavity: report of a rare case and literature review. Braz Dent J. 2019;30:185-190.
37. Smith MH, Bhattacharyya I, Cohen DM, Hinze SR, Islam MN. Glomus tumor: a comprehensive review of the clinical and histopathologic features with report of two intraoral cases. *Oral Surg Oral Med Oral Pathol Oral Radiol*. 2019;127:62-70.

38. Zou H, Song L, Jia M, Wang L, Sun Y. Glomus tumor in the floor of the mouth: a case report and review of the literature. *World J Surg Oncol*. 2018;16:201.

39. Rajendran S, Henderson AH, Gillett S. Rare glomangiosarcoma of the tongue. *BMJ Case Rep*. 2018;2018:bcr2017223268.

40. Carroll RE, Berman AT. Glomus tumors of the hand: review of the literature and report on twenty-eight cases. *J Bone Joint Surg Am*. 1972;54:691-703.

41. Mravic M, LaChaud G, Nguyen A, Scott MA, Dry SM, James AW. Clinical and histopathological diagnosis of glomus tumor: an institutional experience of 138 cases. *Int J Surg Pathol*. 2015;23:181-188.

42. Fletcher CDM, Krishnan UK, Fredrik M; World Health Organization; International Agency for Research on Cancer. *Pathology and Genetics of Tumours of Soft Tissue and Bone*. IARC Press; 2002:136-137.

43. Grover C, Khurana A, Jain R, Rathi V. Transungual surgical excision of subungual glomus tumour. *J Cutan Aesthet Surg*. 2013;6:196-203.

44. Monaghan L, Clark S. Glomus tumor: an unusual cause of a lump in the upper lip. *J Case Rep Images Dent*. 2017;3:36-39.