Intraoral malignant glomus tumor

Satheesh Chandran1, Arun Elangovan2, Saranya Vijayakumar3, K. Sai Sarath Kumar4, Departments of 1Oral and Maxillofacial Surgery, 2Pediatric Dentistry, Madha Dental College and Hospital, 3Department of Oral and Maxillofacial Pathology, Adhiparasakthi Dental College and Hospital, 4Department of Pediatric and Preventive Dentistry, Sathyabama Dental College and Hospital, Chennai, Tamil Nadu, India

Abstract

Glomus tumors are uncommon, benign solitary tumors derived from the glomus apparatus. We report here a case of a malignant glomus tumor in an 8-year-old child presenting as a multilocular ill-defined radiolucency of the mandible. The lesion microscopically showed sheets of round basophilic cells with high nuclear-cytoplasmic ratio, indistinct cell boundaries, nuclear hyperchromatism and nuclear pleomorphism. Immunohistochemically, the tumor was positive for vimentin and smooth muscle actin.

Keywords: Child, glomus tumor, malignant, mandible, oral, pedodontia, vimentin

INTRODUCTION

The glomus apparatus was identified in 1862 by Sucquet and described in detail by Hoyer in 1877. It is an arteriovenous anastomosis located in the stratum reticularis of the dermis predominantly on the palm and digits of the hand and the ventral surface of the feet.[1] It is involved in thermal regulation. The glomus tumor derived from the glomus apparatus accounts for <2% of soft tissue tumors.[2] It is a rare tumor that usually is seen in distal extremities. Other less common sites of involvement include the nasal cavity, middle ear, stomach, bone, lung and rarely oral cavity (0.6%).[3,4]

We present here a rare case of an intraoral malignant glomus tumor in the mandible and review the literature concerning the glomus tumor of the oral cavity.

CASE REPORT

An 8-year-old girl presented to the Department of Pedodontics, Ragas Dental College and Hospital, with an intraoral growth in the alveolar region of the right body of the mandible of 1-year duration with a history of a gradual increase to the present size of 3 × 3 cm [Figure 1]. The sessile growth was soft in consistency and nontender on palpation. Mucosa over the swelling was smooth and not ulcerated. The first molar and deciduous canine and first molar on the affected side were mobile. The patient gave a history of incisional biopsy done a month earlier, reported as “insufficient tissue for diagnosis.” Orthopantomogram revealed an ill-defined multilocular radiolucency in the right body of the mandible measuring 3.5 × 1.5 cm in size and extending from the apical end of 83 to the mesial aspect of the 47 permanent tooth bud. Forty-six and 84 exhibited root resorption. The permanent tooth buds 44 and 45

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were within the radiolucency [Figure 2]. The provisional diagnosis was ameloblastic fibrosarcoma.

An excisional biopsy was done. The gross specimen submitted included hard and soft tissue and measured 2 × 4.5 × 2 cm in size [Figure 3].

Histopathological examination showed uniform, monomorphic round blue cells arranged in sheets and cords. The cells were basophilic with a high nuclear-cytoplasmic ratio, indistinct cell boundaries [Figure 4a], foci of nuclear hyperchromatism, nuclear pleomorphism and mitotic figures (9/10HPF) [Figure 4b]. The connective tissue stroma was fibrovascular with numerous dilated capillaries and focal edematous areas. The tumor cells were positive for vimentin and smooth muscle actin (SMA) [Figures 4c and d] and negative for desmin, p63, CD34 and CD45.

The histopathological features were not consistent with the clinical diagnosis of ameloblastic fibrosarcoma because ameloblastic fibrosarcoma shows scattered odontogenic epithelial cell rests, intertwining fibrils in the connective tissue.

**DISCUSSION**

The glomus tumor is a distinct neoplasm of perivascular cells that resemble modified smooth muscle cells seen in the glomus body. The glomus body is most frequently encountered in the subungual region, lateral areas of the digits and the palm where it is involved in thermal regulation. Glomus tumors are usually solitary, painful and well-circumscribed and treated by simple excision. Rarely, they can be multiple.[1] Table 1 lists the cases of oral glomus tumors reported in the literature from 1949 to 2015.

The term glomangioma for the benign tumor was coined by Bailey in 1935. Masson described the occurrence of three histologic patterns –i) angiomatous-most common, ii) solid comprising cellular areas of smooth muscle cells and epithelioid cells and iii) degenerative with hyalinization, edema and mucoid changes in a myxoid stroma. However,
these patterns may be mixed in varying proportions in any glomus tumor.\textsuperscript{[4,20]}

The World Health Organization classifies glomus tumors as glomangioma (prominent vascular component), glomangiomyoma (prominent smooth muscle component) and solid glomus tumor (prominent cellular component).\textsuperscript{[3,28]}

Variants of glomus tumors include (1) Glomangiomas, a benign, diffuse-growing glomus tumor; (2) Symplastic glomus tumor, characterized by marked nuclear atypia (representing a degenerative phenomenon) and no other features of malignancy and (3) Malignant glomus tumor or glomangiosarcoma, which accounts for approximately 1\% of all glomus tumors.\textsuperscript{[1]}

Our present case showed sheets of round cells with basophilic cytoplasm, high nuclear-cytoplasmic ratio, nuclear hyperchromatism, nuclear pleomorphism and mitotic figures (9/10HPF), all suggestive of a malignant glomus tumor

**Histological differential diagnosis included** hemangiopericytoma, myopericytoma, leiomyosarcoma and gastrointestinal stromal tumor (GIST) [Table 2].

### Table 1: Cases of glomus tumor affecting the oral cavity

| Author          | Year | Age/sex | Anatomic location          | IHC profile                                                                 |
|-----------------|------|---------|---------------------------|------------------------------------------------------------------------------|
| Von Langer\textsuperscript{[5]} | 1949 | 52 male | Hard palate               | Vimentin +, Factor VIII - , CD45 - , A-BGA - , cytokeratin -                  |
| King\textsuperscript{[6]}       | 1954 | 32 male | Gingiva                   | Vimentin +, Factor VIII - , CD45 - , A-BGA - , cytokeratin -                  |
| Kirschner and Strassburg\textsuperscript{[7]} | 1962 | 56 male | Gingiva/alveolar mucosa   | Vimentin +, Factor VIII - , CD45 - , A-BGA - , cytokeratin -                  |
| Grande and D’Angelo\textsuperscript{[8]} | 1962 | 42 male | Hard palate               | Vimentin +, Factor VIII - , CD45 - , A-BGA - , cytokeratin -                  |
| Franke\textsuperscript{[9]}     | 1965 | 13 male | Buccal mucosa             | Vimentin +, Factor VIII - , CD45 - , A-BGA - , cytokeratin -                  |
| Harris and Griffin\textsuperscript{[10]} | 1965 | 35 female | Periodontium/gingiva | Vimentin +, Factor VIII - , CD45 - , A-BGA - , cytokeratin -                  |
| Sidhu and Subherwal\textsuperscript{[11]} | 1967 | 10 female | Hard palate              | Vimentin +, Factor VIII - , CD45 - , A-BGA - , cytokeratin -                  |
| Charles (multiple lesions)\textsuperscript{[12]} | 1976 | 17 female | Hard palate              | Vimentin +, Factor VIII - , CD45 - , A-BGA - , cytokeratin -                  |
| Sato et al.\textsuperscript{[13]} | 1979 | 29 male | Tongue                    | Vimentin +, Factor VIII - , CD45 - , A-BGA - , cytokeratin -                  |
| Tajima et al.\textsuperscript{[14]} | 1981 | 63 female | Tongue                   | Vimentin +, Factor VIII - , CD45 - , A-BGA - , cytokeratin -                  |
| Saku et al.\textsuperscript{[15]} | 1985 | 45 male | Buccal mucosa             | Vimentin +, Factor VIII - , CD45 - , A-BGA - , cytokeratin -                  |
| Ficarra et al.\textsuperscript{[16]} | 1986 | 51 female | Upper lip                | Vimentin +, Factor VIII - , CD45 - , A-BGA - , cytokeratin -                  |
| Moody et al.\textsuperscript{[17]} | 1986 | 65 female | Upper lip                | Vimentin +, Factor VIII - , CD45 - , A-BGA - , cytokeratin -                  |
| Stajcić and Bojić\textsuperscript{[18]} | 1987 | 55 male | Tongue                    | Vimentin +, Factor VIII - , CD45 - , A-BGA - , cytokeratin -                  |
| Geraghty et al.\textsuperscript{[19]} | 1992 | 71 male | Hard palate               | Vimentin +, Factor VIII - , CD45 - , A-BGA - , cytokeratin -                  |
| Kusama et al.\textsuperscript{[20]} | 1995 | 57 male | Upper lip                 | Vimentin +, Factor VIII - , CD45 - , A-BGA - , cytokeratin -                  |
| Sakashita et al.\textsuperscript{[21]} | 1997 | 54 male | Upper lip                 | Vimentin +, Factor VIII - , CD45 - , A-BGA - , cytokeratin -                  |
| Yu et al. (multiple lesions)\textsuperscript{[22]} | 2000 | 54 female | Left mandibular area, lip, anterior buccal mucosa | Vimentin +, Factor VIII - , CD45 - , A-BGA - , cytokeratin -                  |
| Kessaris et al.\textsuperscript{[23]} | 2001 | 46 female | Hard palate              | Vimentin +, Factor VIII - , CD45 - , A-BGA - , cytokeratin -                  |
| Rallis et al.\textsuperscript{[24]} | 2004 | 85 female | Upper lip                 | Vimentin +, Factor VIII - , CD45 - , A-BGA - , cytokeratin -                  |
| Lanza et al.\textsuperscript{[25]} | 2005 | 65 male | Lower lip                 | Vimentin +, Factor VIII - , CD45 - , A-BGA - , cytokeratin -                  |
| Boros et al.\textsuperscript{[26]} | 2010 | 34 male | Lower lip                 | Vimentin +, Factor VIII - , CD45 - , A-BGA - , cytokeratin -                  |
| Per Durand III et al.\textsuperscript{[24]} | 2010 | 11 female | Lower lip                | Vimentin +, Factor VIII - , CD45 - , A-BGA - , cytokeratin -                  |
| Biswas et al.\textsuperscript{[27]} | 2014 | 38 female | The floor of the mouth   | Vimentin +, Factor VIII - , CD45 - , A-BGA - , cytokeratin -                  |
| Mohan et al.\textsuperscript{[28]} | 2015 | 15 female | Upper lip                | Vimentin +, Factor VIII - , CD45 - , A-BGA - , cytokeratin -                  |

IHC: Immunohistochemical, SMA: Smooth muscle actin, MSA: Muscle-specific actin

### Table 2: Differential diagnosis

| Differential diagnosis | Clinical features | Histopathology | IHC profile |
|------------------------|-------------------|----------------|-------------|
| Hemangiopericytoma     | Benign tumor      | Ovoid to spindle cells | CD34 +, SMA - , SMA +, CD34 - , desmin - |
|                        |                   | Spindle-shaped cells |             |
| Myopericytoma          | Benign neoplasm of pericytes | Spindle to round cells | SMA +, CD34 - , desmin - |
| Leiomysarcoma          | Malignant Smooth muscle tumor | Spindle cells arranged in fasciitis | SMA +, CD34 - , c-KIT +, CD34 - |
| GIST                   | Neoplasm in the gastrointestinal tract |         |             |

IHC: Immunohistochemical, SMA: Smooth muscle actin, MSA: Muscle-specific actin, GIST: Gastrointestinal stromal tumor

Hemangiopericytoma is a benign tumor, clinically usually large and situated deep in the connective tissue. Histopathologically, the neoplastic cells of hemangiopericytoma have ovoid to spindle morphology and immunohistochemically show positivity for CD34 and negativity for SMA. Immunohistochemically, our present case was negative for CD34 and positive for SMA.

Myopericytoma is a tumor of neoplastic pericytes with smooth muscle differentiation around vascular channels. The neoplastic cells of myopericytoma are spindle, while
in the present case, the neoplastic cells were round to oval. Myopericytoma is positive for SMA and CD34 as was our present case.

Leiomyosarcoma is a malignant tumor of smooth muscles. The neoplastic cells of a leiomyosarcoma have a spindle to round morphology, whereas neoplastic cells of the glomus tumor are round to oval in morphology. Leiomyosarcoma is positive for desmin which was negative in the present case.

GIST is a mesenchymal neoplasm that arises in the gastrointestinal tract. In pediatric patients, GIST occurs as a component of Carney's triad (gastric GIST, extraadrenal paraganglioma and pulmonary chondroma). GIST has spindle cells and shows immunohistochemistry (IHC) positivity for c-KIT and CD34. The present case was in a pediatric patient, however, the cells were round and were negative for CD34.

The clinical, histopathological features (round cells, cellular and nuclear pleomorphism and mitotic figures) and IHC features (vimentin and SMA positivity) led to the diagnosis of malignant glomus tumor.

**CONCLUSION**

- Malignant glomus tumor is one of the rare sarcomas in the oral cavity
- It is a high-grade sarcoma and should be treated immediately to avoid metastasis
- It is necessary to consider malignant glomus tumor as one of the differential diagnoses in round cell sarcoma histopathologically
- To our knowledge, this is the first case of malignant glomus tumor reported in the oral cavity.

**Declaration of patient consent**

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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**Conflicts of interest**

There are no conflicts of interest.

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