

Intraoral malignant glomus tumor

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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

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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 $2 \times 4.5 \times 2$ 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,



Figure 1: Intraoral growth in the alveolar region body of the mandible



Figure 2: Orthopantomogram revealed an ill-defined multilocular radiolucency

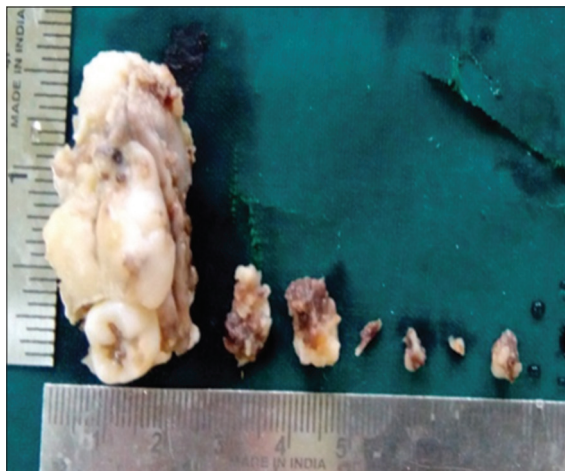


Figure 3: Gross specimen

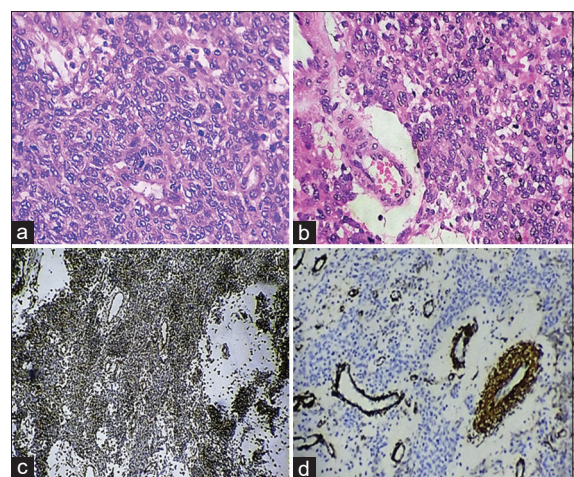


Figure 4: (a) Sheets of round cells with basophilic cytoplasm, high nuclear-cytoplasmic ratio and indistinct cell boundaries (H and E, $\times 40$). (b) The presence of mitotic figures (H and E, $\times 40$). (c) The tumor cells show diffuse positivity for vimentin (immunohistochemistry, $\times 10$). (d) The tumor cells show focal positivity for smooth muscle actin (immunohistochemistry, $\times 10$)

Table 1: Cases of glomus tumor affecting the oral cavity

Author	Year	Age/sex	Anatomic location	IHC profile
Von Langer ^[5]	1949	52 male	Hard palate	Not available
King ^[6]	1954	32 male	Gingiva	Not available
Kirschner and Strassburg ^[7]	1962	56 male	Gingiva/alveolar mucosa	Not available
Grande and D'Angelo ^[8]	1962	42 male	Hard palate	Not available
Frankel ^[9]	1965	13 male	Buccal mucosa	Not available
Harris and Griffin ^[10]	1965	35 female	Periodontium/gingiva	Not available
Sidhu and Subherwal ^[11]	1967	10 female	Hard palate	Not available
Charles (multiple lesions) ^[12]	1976	17 female	Hard palate	Not available
Sato et al. ^[13]	1979	29 male	Tongue	Not available
Tajjima et al. ^[14]	1981	63 female	Tongue	Not available
Saku et al. ^[15]	1985	45 male	Buccal mucosa	Actin +, smooth muscle myosin +
Ficarra et al. ^[16]	1986	51 female	Upper lip	Not available
Moody et al. ^[17]	1986	65 female	Upper lip	Vimentin +, Factor VIII -, CD45 -, A-BGA -, cytokeratin -
Stajčić and Bojčić ^[18]	1987	55 male	Tongue	Not available
Geraghty et al. ^[19]	1992	71 male	Hard palate	Alpha actin -, neuron-specific enolase -, Chromogranin -, desmin -
Kusama et al. ^[20]	1995	57 male	Upper lip	S-100 +, actin +, desmin +, vimentin +, Factor VIII -
Sakashita et al. ^[21]	1997	54 male	Upper lip	Vimentin +, SMA +, Factor VIII -
Yu et al. (multiple lesions) ^[22]	2000	54 female	Left mandibular area, lip, anterior buccal mucosa	SMA +, S-100 -
Kessarar et al. ^[23]	2001	46 female	Hard palate	Vimentin +, S-100 +, actin -, desmin -, chromogranin -, neuron-specific enolase -, epithelial membrane antigen -, cytokeratin -, Factor VIII -
Rallis et al. ^[24]	2004	85 female	Upper lip	SMA +, MSA+Vimentin +, desmin -, AE1/3 -, S-100 - Epithelial membrane antigen - Neuron-specific enolase - CD3, CD31, CD34, CD45, CD20 - Cytokeratin -, Leu 7 -
Lanza et al. ^[25]	2005	65 male	Lower lip	Not available
Boros et al. ^[3]	2010	34 male	Lower lip	SMA +, MSA+S-100 +, keratin - Epithelial membrane antigen - CD34 -, CD31 -, chromogranin -
Per Durand III et al. ^[26]	2010	11 female	Lower lip	Epithelial membrane antigen, S-100 SMA+, vimentin +, pan cytokeratin
Biswas et al. ^[4]	2014	38 male	The floor of the mouth	Not available
Mohan et al. ^[27]	2015	15 female	Upper lip	Not available

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

these patterns may be mixed in varying proportions in any glomus tumor.^[4,25]

The World Health Organization classifies glomus tumors as glomangioma (prominent vascular component), glomangiomyoma (prominent smooth muscle component) and solid glomus tumor (prominent cellular component).^[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.^[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 2: Differential diagnosis

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

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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