Frontiers in Dentistry

Intraoral Cellular Schwannoma Involving Maxillary Gingiva: A Rare Case Report

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Article Info

Article type:
Case Report

Article History:
Received: 02 Sep 2022
Accepted: 26 Mar 2023
Published: 25 Sep 2023

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A B S T R A C T

Oral schwannomas (OSs) are uncommon benign nerve sheath tumors accounting for 1% of all schwannomas and may arise from either soft tissue or bone. Cellular schwannoma is a rare histological variant of schwannoma which is characterized by increased cellularity. The most common intraoral site of occurrence is the tongue followed by the floor of the mouth and palate. Here, we are reporting a rare case of intraoral cellular schwannoma involving both facial and palatal gingiva of the right maxilla in a young Indian male patient.

Keywords: Gingiva; Mouth; Nerve Sheath Neoplasms; Neurilemmoma

➢ **Cite this article as:** Bagul S, Chandan S, Pandey ND, Choudhary SH. Intraoral Cellular Schwannoma Involving Maxillary Gingiva: A Rare Case Report. *Front Dent*. 2023:20:36.

INTRODUCTION

Schwannoma, as the name suggest, is a benign neurogenic tumor originating from Schwann cells of the nerve sheath [1,2]. Other nomenclature used for this entity includes neurilemmoma, neurinoma and Schwann cell tumor [2]. Unlike classical schwannoma, ancient schwannoma (AS) is a rare long-standing variant of this tumor which shows degenerative changes [3,4], whereas cellular schwannoma (CS), which is an even more rare variant, differs by its increased cellularity, nuclear pleomorphism, hyper-chromatism, lack of Verocay bodies, and frequently higher mitotic activity. Although some cases of CS have been confused histologically with malignancies, only a few have recurred, while none have metastasized [5]. Schwannomas may arise at any age but they are often known to occur between 20-50 years with a slight female predilection [2]. These benign tumors most often affect the head and neck region with an incidence of about 25-45%. However, intraoral occurrence of schwannoma is rare accounting for merely 1% of the total incidence [6]. Tongue is the most commonly affected intraoral site followed by palate, floor of mouth, buccal mucosa and gingiva. Cellular schwannoma arising in the oral cavity is exceedingly rare and only 4 cases have been reported in the literature so far [5-8]. We hereby report a case of cellular schwannoma involving the anterior maxillary gingiva in a 27-year-old male patient.

CASE REPORT

A 27-year-old male reported to the department of

Oral and Maxillofacial Surgery with the chief complaint of gingival overgrowth in the region of his upper right front teeth. History revealed that the swelling was initially smaller in size and had increased slowly over the period of 6 months to its present size. The patient also gave a history of pain and bleeding from the same region while brushing, which had started 1 month ago. There was no history of pus discharge from the area. Past medical, dental and family history was non-contributory. On extra-oral examination, there was facial asymmetry due to the presence of bulging on the right side of the upper lip in association with the intraoral gingival over-growth. Intraoral examination revealed gingival overgrowth in the maxillary right anterior teeth region extending mesiodistally from the maxillary right central incisor (8) to the maxillary right first premolar (5). It encompassed the entire facial aspect, extending from the muco-buccal fold to the palatal gingiva. The overgrowth partially covered the crown of the right central incisor (8) and half of the crown length of the right lateral incisor (7). On the palatal aspect, the overgrowth extended 1.5 cm posteriorly from the free gingival margin and was associated with the maxillary lateral incisor (7), canine (6) and first premolar (5) teeth. The overlying mucosa appeared reddish in color in certain areas of the overgrowth. (Figure 1)

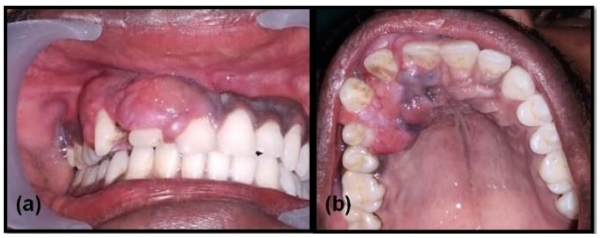

Fig. 1. Intraoral clinical picture of the lesion (a) on facial aspect; (b) on palatal aspect

On palpation, the overgrowth was tender and soft to firm in consistency and was attached to the underlying tissue. There was bleeding on probing and grade III mobility in the maxillary lateral incisor and canine. Based on the history and clinical examination, a provisional diagnosis of pyogenic granuloma associated with the maxillary right anterior teeth region was made. Differential diagnosis included peripheral giant cell granuloma, localized periodontitis, peripheral ossifying fibroma, and fibrous hyperplasia. All routine hematological investigations revealed normal values. Intraoral periapical radiographs showed a well-defined radiolucency involving 8, 7, 6 regions. Tooth number 7 was associated with severe bone loss and horizontal bone loss was seen around 6 (Figure 2).

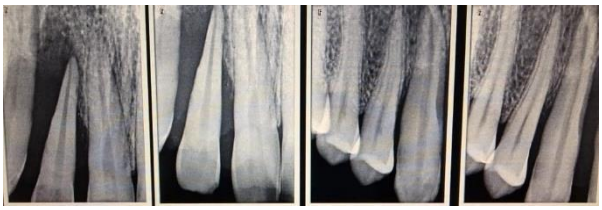

Fig. 2. Intraoral periapical radiograph showing the level of bone loss in the right maxillary quadrant from 5-8 region

An incisional biopsy was taken from the border of the lesion and the specimen was sent for histopathologic evaluation, which revealed the presence of fibrovascular connective tissue exhibiting proliferation of nerve fiber bundles and spindle-shaped cells with wavy nuclei arranged in a palisading manner. Cellular and nuclear pleomorphism was seen in focal areas (Figure 3). Antoni A and Antoni B-type cell patterns were also observed. Histopathological examination was suggestive of a benign yet aggressive neural tumor.

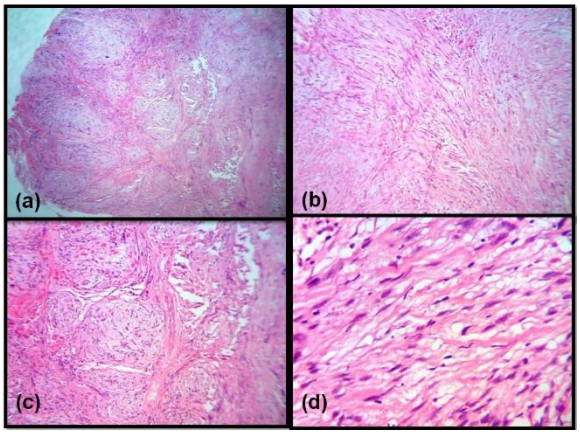

Fig. 3. Histopathological H&E image showing (a) fibrovascular connective tissue exhibiting proliferation of spindle-shaped cells; (b) hyper-cellular Antoni A pattern; (c) marked cellularity with fascicles of spindle cells, displaying short intersecting fascicles and whorls of Schwann cells;(d) spindle shaped cells with wavy nuclei along with cellular and nuclear pleomorphism

In order to confirm the diagnosis, immune-histochemical staining was performed using S-100, SMA, vimentin and Ki-67. The tissue showed 100% positive immunohistochemical reactivity with S100 protein (Figure 4a), thus confirming the neural origin. Ki 67 (Figure 4b) showed 60% positivity in hot spot areas suggesting increased DNA proliferation and aggressive behavior of this tumor. Vimentin (Figure 4c) also showed positive reactivity in the connective tissue, while p53 (Figure 4d) was weakly positive, with only 10% of cells showing immunoreactivity.

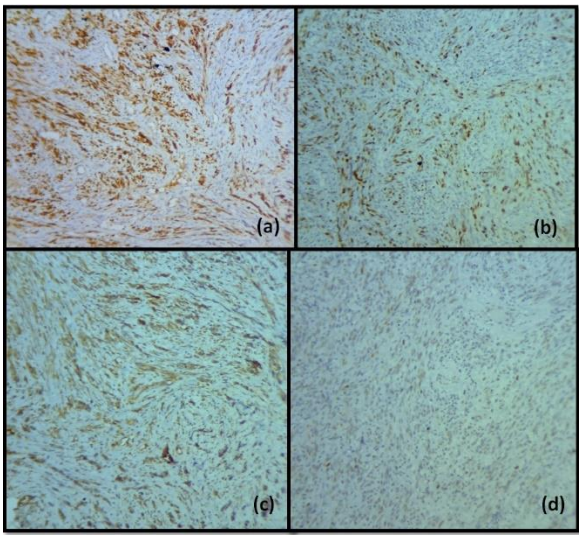

Fig. 4. Immunohistochemical images showing reactivity of cells for (a) S-100 protein, (b) Ki67, (c) Vimentin and (d) p53.

Thus, a final diagnosis of cellular schwannoma was made. The treatment plan consisted of surgical excision of the lesion along with extraction of the associated teeth, followed by intraoral prosthesis post-healing. The prognosis was predicted to be poor. The situation and treatment plan were explained in detail to the patient and a written informed consent was obtained prior to surgery. Wide local excision with a soft tissue margin of 5mm around the lesion was performed under the infraorbital nerve block, nasopalatine and greater palatine nerve blocks. Labial infiltration was also considered in order to hasten hemostasis. Extraction of 7 and 6 was performed due to anticipated poor prognosis. (Figure 5) Vulcanite bur was used to smoothen the sharp bony edges under irrigation. Hemostasis was achieved and since primary closure of the wound was not possible because of well-separated margins, a gauze soaked in iodine solution was placed over the bare bone and secured with 3-0 silk suture for the first two days and was changed subsequently.

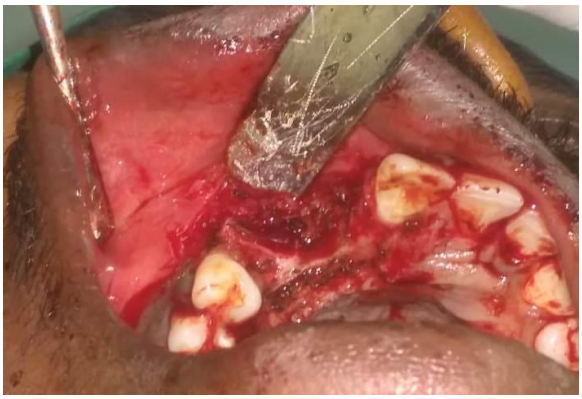

Fig. 5. Intra-operative picture after excision of the lesion and extraction of 7 and 6

Secondary healing was achieved by 3 weeks and the wound healed over a period of one month. The lesion was removed as one large mass with few small pieces. (Figure 6)

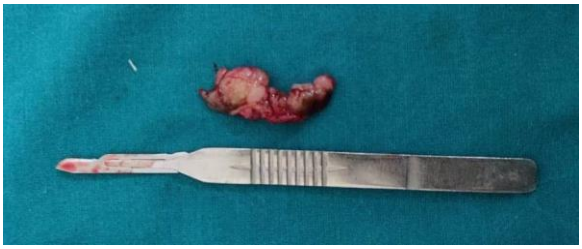

Fig. 6. Excised gingival mass

Post-operative healing was uneventful and the patient is under regular follow-up by telephone and there has been no sign of recurrence within the past year.

DISCUSSION

Schwannoma, also known as neurilemmoma, is an uncommon, slow growing, solitary, benign nerve sheath tumor that originates from peripheral, cranial or autonomic nerves containing schwann cells [9] Histologically, there are seven variants of schwannoma described in the literature which includes classical (Verocay), plexiform, cellular, cranial nerve, melanotic, degenerated (ancient), and granular cell schwannoma [2,10] Verocay, in 1910 was the

first to describe schwannoma microscopically and called it "Neurinoma," while Arthur Stout, in 1935, coined the term neurilemmoma [7,10]. Between 25%–45% of schwannomas are located in the head and neck region, with the eighth cranial nerve being most commonly affected. Only 1% of the schwannoms are known to occur intraorally, with tongue being the commonest site [7,9]. To the best of our knowledge, so far only four cases of intraoral cellular schwannoma have been reported in literature. The first case of CS was reported in 1996 involving the left mandibular canal region [5], second was reported in 2002 involving the oral mucosa of a Japanese male patient [6], the third case was reported as multiple swellings involving the right quadrant of both maxilla and mandible in 2012 [7] and the fourth case was found in 2022 involving the buccal mucosa on the left side [8]. The present case is the fifth reported case of intraoral CS with an unusual site of presentation (maxillary anterior gingiva) that makes it unique.

CS is known to affect individuals in their second to fifth decades of life with a slight female predilection with a female to male ratio of 1.6:1 [7]. Out of all the four cases of intraoral CS patients reported previously, two were in their 3rd decade and two were in their 4th decade of life. Also, out of four, two were male and two were female patients [5-8]. In the present case, the patient was a 27 years old male.

Schwannoma can be either central (bone) or peripheral (soft tissue) [7]. The etiology of schwannoma is still unknown but it is believed to originate from the proliferation of Schwann cells in the perineurium resulting in displacement and compression of the adjacent structures [7,11]. Schwannoma can occur either in an encapsulated form, which is more common or in a pedunculated form, where it resembles a fibroma. Clinically, it often presents as a painless slow growing, solitary, exophytic, non-indurated, well circumscribed lesion, with a smooth surface [9]. The very first case in intraoral CS reported in 1996 was an incidental finding [5]. Also all the previously reported cases of intraoral CS were painless in nature. However, in the present case, the lesion presented as a diffuse gingival overgrowth in the right maxillary anterior region associated with pain and bleeding on brushing, which is a rare clinical presentation. If multiple lesions are present, the term "Schwannomatosis" is used wherein cranial, spinal or peripheral nerves can be involved. Schwannomatosis is often observed with or without neurofibromatosis or Von Recklinghausen's disease [7].

Depending upon the clinical presentation of the lesion, various differential diagnoses are considered including fibroma, granular cell tumor, pyogenic granuloma, giant cell lesion, fibrous hyperplasia, traumatic fibroma, mucocele, minor salivary gland tumors, and rarely, malignant tumors, such as squamous cell carcinoma and sarcomas [2,9]. Radiographically, schwannomas of jaw appears as well-demarcated, unilocular radiolucencies within a thin sclerotic border. Other findings like external root resorption, cortical thinning, cortical expansion, and peripheral scalloping are also evident in some cases. As diagnostic tools, various other investigations such as ultrasonography, computed tomography, and magnetic resonance imaging often proves helpful for estimation of tumor margins as well as the local invasion in surrounding structures [7,12].

On histological evaluation, classic schwannoma is marked by the presence of a mixture of Antoni A and Antoni B patterns. Antoni A is comprised of hypercellular spindle cells arranged in a palisading fashion with acellular zones alternating in between, the Verocay bodies, while Antoni B pattern is referred to the hypocellular, loosely textured areas [13]. Cellular schwannoma is distinguished by the presence of Antoni A pattern almost entirely and the absence of Verocay bodies, nuclear pleomorphism and hyperchromatism, and frequently higher mitotic activity [5,13], as was observed in the present case. The presence of Schwann cells in whorls, a peritumoral capsule, macrophage-rich infiltrates, subcapsular lymphocytes, and the absence of fascicles distinguishes the cellular schwannoma from malignant peripheral nerve sheath tumor which shows the presence of perivascular hyper-

cellularity, vascular invasion, and necrosis. Although CS and classic schwannoma occurs in a similar age group, CS usually affects the retroperitoneum region and often display histopathologic findings resembling that of sarcoma or sarcomatoid carcinoma [13]. In the present case, p53 immunohistochemical staining was performed to rule out malignancy. On immunohistochemistry, CS are known to be positive for S100 which reflects the neural nature of the lesion [7,9], as was performed in the present case that yielded positive results. Ki-67 labeling indices ≥20% is indicative of malignant peripheral nerve sheath tumor with 87% sensitivity and 96% specificity which was not observed in the present case. This case demonstrated 60% immunoreactivity with Ki-67 in hot spot areas whereas the overall expression was ≤ 20%. The final diagnosis of CS was confirmed based on the routine hematoxylin and eosin staining along with immunohistochemical evaluation of S-100, Ki67, vimentin and p53. The diagnosis of CS is based entirely on histopathological and immune-histochemical evaluations [13].

Local surgical excision is the recommended treatment for CS. In case of non-encapsulated schwannoma, a margin of normal tissue and careful separation from the involved nerve becomes mandatory to preserve normal functioning post operatively [8]. However, in the present case, nerve involvement was not evident and hence the mass was completely excised. Schwannomas are known to have a good prognosis since they rarely recur and malignant transformation is also rare [9].

CONCLUSION

Oral CS is an uncommon tumor and it rarely affects gingiva. This is the forth reported case of intraoral CS in literature. It is essential that every painless solitary swelling of oral cavity should be meticulously examined and various differential diagnosis should be considered. This article suggests that it is important for the dental clinicians to consider schwannoma in the differential diagnosis of any gingival mass, especially when other causative factors of gingival enlargement are ruled out. Since the final diagnosis of schwannoma can be confirmed only after histopathological evaluation, histopathological examination of every excised mass should be regularly practiced.

CONFILICT OF INTEREST STATEMENT

None declared.

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