A Rare Report of Two Cases: Ancient Schwannoma of Infratemporal Fossa and Verocay Schwannoma of Buccal Mucosa

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Abstract

Schwannoma is a benign tumor that originates from Schwann cells of the peripheral nerve. Occurring as a common tumor in the head and neck region, its intraoral presentation is rare which accounts for only 1%. They are solitary, slow-growing, smooth-surfaced, usually asymptomatic, and encapsulated tumor. This paper comprises two case reports, both of which presented with an intraoral soft tissue swelling which turned out to be schwannoma of buccal mucosa and infratemporal fossa with extension into the oral cavity, respectively. The treatment was done with complete surgical excision. Soft tissue tumors of the oral cavity can present with similar clinical feature, which can make it difficult to distinguish one lesion from the other. Thus, final diagnosis for such lesions can only be rendered based on complete analysis of clinical, radiological, surgical, and histopathological findings.

Keywords: Buccal mucosa, infratemporal fossa, neurilemmoma, schwannoma

Introduction

Schwannomas are benign, slow-growing, epineurium-encapsulated neoplasms arising from Schwann cells that comprise myelin sheaths surrounding peripheral nerves. Although 25%–48% of all schwannomas occur in the head and neck region, the development of this tumor in the oral cavity is quite uncommon representing only 1% of all head and neck region tumors. Schwannomas of the head and neck occur intracranially, mainly at the cerebellar pontine angle, and in peripheral soft tissues, mostly involves the tongue followed by the palate, floor of the mouth, oral mucosa and mandible. The infratemporal fossa is one of the least common anatomical sites for schwannomas. The tumor can arise at any age but is most common in the third and fourth decades of life. It is not certain whether there is a predilection for men, women or it is equally distributed between sexes.

The clinical differential diagnosis could be with any other benign tumors such as fibroma, lipoma, neurofibroma, or salivary glands tumor. The treatment of choice is complete excision and recurrence is rare.

Case Reports

Case 1
A 36-year-old male patient reported with swelling in the right facial region for 1 year. On extraoral examination, a mild diffuse swelling was present on the right midfacial region. His medical history was unremarkable. Intraoral examination revealed a 2 cm × 2 cm, sessile, firm, nontender, nonulcerated mass on the right posterior buccal mucosa [Figure 1a]. There was a history of extraction with respect to 17, 18 due to cheek biting. Ultrasound examination revealed a 30 mm × 23 mm hypoechoic mass in the soft tissue of right cheek, following which fine-needle aspiration cytology was conducted, which was inconclusive.

Contrast-enhanced computed tomography in axial sections showed 3.05 cm × 2.62 cm round, hypodense lesion in the

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How to cite this article: Piyush P, Mohanty S, Ghosh S, Gupta S. A rare report of two cases: Ancient schwannoma of infratemporal fossa and verocay schwannoma of buccal mucosa. Ann Maxillofac Surg 2017;7:140-2.
right retromaxillary/infratemporal region compressing the posterior wall of maxillary sinus and anteromedially bulging into the oral cavity. There was no definite erosion/destruction of the underlying bone [Figure 1b]. As the lesion appeared to be well encapsulated, complete surgical excision was planned. The entire mass was excised in toto [Figure 1c] and was sent for histopathological examination.

The sections showed well-encapsulated highly cellular tumor with spindle-shaped cells. The tumor consisted predominantly Antoni B pattern; bundles of palisading cells were seen with buckled nuclei [Figure 1d]. Thus, a diagnosis of Schwannoma was established. The patient is under regular follow-up, with no recurrence 1 year after surgery.

Case 2
A 23-year-old female patient complained of swelling in the right cheek region for 1 year. It was associated with pain of mild intensity. The patient visited a local dentist and got the extraction of 18 and 38 3 months back after which some relief was obtained but the pain recurred for 2 months. There was no significant medical history.

Extraorally, a diffuse swelling on the right facial region was seen. Intraoral examination revealed a diffuse growth on the right posterior buccal mucosa. On palpation, it was around 1 cm × 0.5 cm, firm, tender, and noncompressible. There was no bleeding or ulceration of the surface [Figure 2a]. Magnetic resonance imaging of the patient in coronal and axial section [Figure 2b and 2c] revealed a 6 cm × 4.5 cm well-defined lobulated heterogeneously enhancing lesion with solid as well as necrotic component in the right infratemporal region. Right zygomatic arch was involved with focal erosion of inner table of ramus of the mandible. There was focal erosion of right maxilla inferiorly.

Incisonal biopsy showed lesional tissue composed of neural cells with spindled nuclei. All lesional cells were positive for S-100 antibody [Figure 2d]. The diagnosis of benign peripheral nerve sheath tumor was made and complete surgical excision was planned. With intraoral approach, incision in right maxillary vestibule and palate was made and osteotomy of the maxilla was done to access and expose the tumor, which was removed completely [Figure 2e]. Histopathological sections showed areas of hyalinization and myxoid change with peripherally placed nuclei. Thick blood vessels with hyalinization and perivascular inflammation were noted with cystic changes.

The overall histo-architecture was consistent with diagnosis of ancient schwannoma [Figure 2f]. The patient evolved satisfactorily with no recurrence 1 year after surgery.

Discussion
Schwannoma (neurilemmoma) is a rare, benign, encapsulated perineural tumor of neuroectodermal origin that is derived from the Schwann cells of the neural sheath. Earlier, Verocay referred to it as a “neuronoma.” Later, Stout used the term, “neurilemmoma,” believing that this tumor originated from the Schwann cells.[8]

It can present itself at any age. However, it is more common between 30 and 50 years of life.[5] William et al. findings showed that in 83% of the cases studied by them, schwannomas presented in males, while for Lucas, there was a greater predilection for females, and for Hatziotis and Asprides, Enzinger and Weiss, there was an equal distribution between

Figure 1: (a) Intraoral picture showing well-defined soft tissue mass in the right posterior buccal mucosa. (b) Contrast-enhanced computed tomography in axial sections showing 3.05 cm × 2.62 cm round, hypodense lesion in the right retromaxillary region. (c) Completely excised tumor mass. (d) Bundles of palisading cells with buckled nuclei (H and E, ×10)

Figure 2: (a) Intraoral swelling in right maxillary vestibule. (b and c) Magnetic resonance imaging in axial and coronal section showing 6 cm × 4.5 cm well-defined lobulated heterogeneously enhancing lesion with solid as well as necrotic component in right infratemporal region. (d) Lesional tissue showing S-100 positivity. (e) Completely excised tumor mass. (f) Areas of hyalinization and myxoid change with peripherally placed nuclei (H and E, ×10). Thick blood vessels with hyalinization and perivascular inflammation were noted with cystic changes and fibrin deposition.
both sexes.\[^6\] In the oral cavity, the lesion is usually present in soft tissues, more commonly the tongue, followed by the palate and buccal mucosa and may have clinical aspects similar to other benign lesions such as mucocele, fibromas, lipomas, and benign salivary gland tumors.\[^9\] Ancient schwannoma usually present as submucosal swellings with a higher female predilection.\[^4\] The presenting lesion can be a primary in the infratemporal fossa with extension into the oral cavity or vice versa. Similar finding was present in our report, where primary site was buccal mucosa in the first case and infratemporal fossa in the second case. The female gender and long-standing history were also consistent with the literature.

In 1985, Erlandson\[^{10}\] classified schwannomas into seven subtypes: classical (Verocay), cellular, plexiform, cranial nerve, melanotic, degenerated (ancient), and granular cell schwannoma.

Schwannomas are designated as ancient when there is either cystic or fatty degeneration, focal accumulations of hyaline material, thick capsule, and infiltration of histiocytes, siderophages, and hyperchromatism. These histopathological features were attributed to the long duration of the schwannoma as consistent with the second case.\[^4\]

The treatment of choice is pericapsular excision. The schwannoma should be extirpated in its entirety to avoid tumor recurrence even if the nerve of origin cannot be preserved.\[^9\] The prognosis is very good and malignant transformation is rare although some authors have mentioned a malignant transformation rate of 8%–13.9%.\[^9\]

**Conclusion**

The low incidence of schwannoma often leads to omission of the tumor in the differential diagnosis of soft tissue tumors of the oral cavity. As the clinical and radiological pictures are not distinctive, final diagnosis should be made after histopathological examination and in some cases after immunohistochemistry analysis. Even, tumors of the infratemporal fossa can be completely excised through an intraoral approach. The treatment of choice is complete surgical excision as recurrences and malignant transformations are exceedingly rare.

**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.

**Financial support and sponsorship**

Nil.

**Conflicts of interest**

There are no conflicts of interest.

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