Parapharyngeal Space Schwannoma: Report of a Case with Brief Review
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Abstract
Schwannomas are benign, encapsulated, slow-growing tumors originating from Schwann cells of the peripheral nerve sheath. Approximately 25-40% of cases occur in the head and neck region, of which one percent has been reported in the oral cavity. They are usually asymptomatic and malignant transformation is rare. Few cases of intraoral schwannoma occurring on the tongue, the floor of the mouth, palate, gingiva, vestibule, lips, salivary glands and the mental nerve region have been reported in the medical literature. This paper reports a rare presentation of an intraoral schwannoma located in the parapharyngeal space arising from the parotid gland in a 31-year-old female patient.

Keywords: Neurilemmoma; Oral; Pharyngeal; Peripheral Nerve Sheath Tumors; Schwann cells; Schwannoma.

Introduction
The schwannomas, also called Neurilemmomas, are typically benign, slow growing, and asymptomatic tumors which are formed by proliferating Schwann cells encompassing peripheral motor and sensory nerves. cultivation of all schwannomas originate in the head and neck region. They are reported to occur in the face, scalp, intracranial cavity, orbit, nasal and oral cavities, parapharyngeal space, middle ear, mastoid, larynx, and medial and lateral regions of the neck. Neurilemmoma was first described by Verocay in 1910. He called it "Neurinoma" then. In 1935, the term 'Neurilemmoma' was coined by Stout. Schwannoma occur in the oral cavity at tongue, floor of the mouth, palate, gingiva, vestibule, lips, salivary glands and mental nerve region. Histopathologically, five schwannoma variants have been described: classical, plexiform, cellular, epithelioid, and ancient schwannomas.

Case Report
A 31-year-old female reported to the Department of Oral and Maxillofacial Surgery with a chief complaint of a painless swelling in her left lower jaw which she had noticed since one month. There was no significant family history and medical history associated. The clinical examination revealed a swelling behind the angle of the mandible on the left side. The swelling measured 3.2 X 2.0 cm in size and was firm in consistency. Lesion was pulsatile and the overlying skin appeared normal. On intraoral examination, the swelling was seen behind the tonsillar fold and the uvula appeared to be shifted to the left.

Fine needle aspiration revealed hemorrhagic fluid and the findings were not contributory. Magnetic Resonance Imaging (MRI) revealed a large oval, homogeneously well-encapsulated mass predominantly in the left parapharyngeal space (Fig 1a). The mass appeared to indent and displace the left internal carotid artery (ICA) anteriorly. The lesion was seen to indent the pharyngeal mucosal space medially. Laterally the lesion appeared to abut from the sternocleidomastoid muscle while anteriorly it abuts the soft tissue in the floor of the mouth. Posteriorly the lesion was seen to be limited by the prevertebral space. Central hypointense areas as on T1 and T2W images suggested necrosis. The patient underwent complete removal of the tumor under general anesthesia. The submandibular approach was the preferred approach for the surgery (Fig 1b).

The gross specimen was lobulated, solid & encapsulated, approximately 4.5 x 3.5 x 2.0cm in size. The mass was yellowish to grayish in color, homogeneous, and soft to firm in consistency with focal areas of hemorrhage (Fig 1c). The lymph node specimen was a single grayish soft piece of tissue measuring 2.2 x 1.0 x 0.5cm. Histopathologic examination of the...
hematoxylin and eosin stained section showed that the lesion was well encapsulated (Fig 1d). The lesional tissue was composed of well organized areas of Antoni A type tissue including bundles of spindle cells with long basophilic nuclei palisaded around acellular eosinophilic areas (Verocay bodies). Antoni B tissue with randomly arranged spindle cells, within a loose myxomatous stroma was also seen. The cells did not exhibit any pleomorphism or hyperchromatism. The entire tumor mass was quite cellular and not many mitotic figures were evident. Diagnosis of schwannoma was given and the lymph node biopsy revealed that there were reactive inflammatory changes in the lymph nodes.

Figure 1: The magnetic resonance imaging revealed a large homogeneously well-encapsulated mass predominantly in the left parapharyngeal space (a). Surgical exposure of tumor by submandibular approach (b) with the gross specimen showing lobulated, solid, encapsulated mass and focal areas of hemorrhage (c). The photomicrograph of hematoxylin and eosin stained section showing the encapsulated lesion composed of well-organized areas of Antoni-A type (Verocay bodies) and Antoni-B type tissue (d).

Discussion
Schwannomas (also referred to as neurilemmomas) are benign encapsulated nerve sheath tumors composed primarily of Schwann cells in a poorly collagenized stroma. It is believed to cause displacement and compression of the adjacent nerve. The parapharyngeal space is located deep within the neck lateral to the pharynx and medial to the ramus of the mandible. In the review of the literature, tumors of the parapharyngeal space are rare, approximately 0.5% of all head and neck tumors. Salivary gland neoplasms are the most common parapharyngeal tumors accounting for 40-50% neoplasms, 20% are of neurogenic origin and enlarged lymph nodes comprise rest 15% to 20%. In the parapharyngeal space, schwannomas most commonly arise from vagus nerve and cervical sympathetic chain. Schwannomas of the parapharyngeal space are usually reported to occur in patients between the age group of 30 and 70 years, there is no sex predilection.

Clinically, schwannoma is a slow growing tumor that frequently produces expansion of
the affected bone and causes swelling. The tumor is solitary with a smooth surface, and a slow asymptomatic growth is evident; although the clinical symptomatology depends on the nerve of origin. Pain and paresthesia may be found in 50% of the patients. These tumors commonly arise in soft tissue of the head and neck region. About 25% to 45% of schwannomas occur in the head and neck and 1% only demonstrates an intraoral origin. Two types are distinguished, central or peripheral Schwannoma, located in bone or in soft tissues respectively. Intraoral schwannomas are rare, particularly in the intraosseous region of the jaw, and less than one percent of tumors occur there. The most common site of occurrence is the mandible.

Imaging techniques such as computed tomography and magnetic resonance are very helpful. Initially, the schwannoma appears as a well-defined tumor. Later with time cystic changes can appear probably associated with mucinous degeneration, hemorrhages and necrosis. In MRI the tumor can present as solid, cystic or mixed. MRI not only determines the tumor but also the nerve from which it develops. The differential diagnosis for a mass found in the parapharyngeal space is wide and can include tumors of the deep lobe of the parotid gland, tumors of minor salivary gland origin, metastatic cervical nodes, paragangliomas, branchial cysts, lymphomas, neurofibromas

In schwannoma, classically two histological patterns are defined, Antoni-A (with hypercellularity) and Antoni-B (with hypocellularity). Antoni-A type is formed by fusiform cells with elongated nuclei arranged in a well-organized palisading pattern. Antoni-A areas with compact aggregates of spindle cells, which are separated by collagen and frequently formed parallel palisading arrays (Verocay bodies). Verocay bodies are formed by two compact rows of well-aligned palisading nuclei with intervening cell processes. Antoni-B type is composed of a smaller number of cells and the spindle cells which are randomly arranged within a loose myxomatous stroma.

In addition to the classic schwannoma, there are several histopathologic variants, including the cellular, plexiform, epithelioid, ancient, and melanotic types. Ackerman and Taylor first found the schwannoma presented with clear areas of hypocellular tissues and attributed the changes to the long standing degenerative changes. Ancient schwannomas tend to be large tumors of long duration and are characterized by degenerative alterations including cystic changes, calcification, hemorrhage, and hyalinization. Macrophages and hemosiderin deposits are common. Although nuclear atypia may be striking, mitotic activity is absent.

The differential diagnosis (by means of histological study) should be made with traumatic neuroma, solitary neurofibroma, granular cell tumor, neurofibromatosis, malignant schwannoma (also called neurogenic sarcoma). Immunohistochemically schwannomas show positivity for S-100, CD 34, and epithelial membrane antigen (EMA), only in the capsule protein.

Although schwannomas are typically benign, they may affect adjacent tissues by expansion with pressure effect. If the schwannoma is completely removed, recurrence rates are extremely low. The treatment of choice is excision. The encapsulated form can be enucleated easily, whereas the non-encapsulated requires normal tissue margins to avoid relapse. If the nerve of origin is visualized, an attempt should be made to separate it carefully to preserve function, although this is sometimes not possible. The prognosis is very good since it does not usually recur, and malignant transformation is rare.

Conclusion
We present here a rare case of intraoral schwannoma. Although they are rare in the oral cavity, the unique nature of presentation imposes a systematic work-up for an accurate diagnosis which includes the histological analysis along with immunohistochemical assay integrated with the clinical data and imaging techniques. Treatment of schwannoma consists of surgical enucleation with periodic follow-up examinations. Recurrence is uncommon and in present case, the patient was followed up for one year with no clinical or radiographic signs of recurrence.

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