Neurilemmoma: A Rare Histopathological Presentation

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Neurilemmomas are benign tumors arising from the Schwann cells of peripheral, cranial, and sympathetic nerves. Neurilemmoma or schwannoma is typically solitary, slow growing, and encapsulated neoplasm that originates in a nerve and is composed of Schwann cells in a collagenous matrix. The tongue is the most common intraoral site, but neurilemmoma of buccal mucosa is seldom reported. This paper reports a rare case of neurilemmoma in 54-year-old female patient in the right buccal mucosa with uncommon histopathological presentation and microscopically confirmed with immunostaining of S-100 protein.

Keywords: Clear cells, Neurilemmoma, Schwann cells, S-100

INTRODUCTION

Neurogenic tumors include benign and malignant variants; benign group consists of neurofibroma, schwannoma, neuroma and perineuroma, and malignant group constitutes malignant peripheral nerve sheath tumors (neurogenic sarcoma, neurofibrosarcoma, and malignant schwannoma).¹ Neurilemmoma is a benign, encapsulated perineural tumor, that originates from the Schwann cells of the nerve sheath derived from the neuroectoderm.² One-fourth of extracranial neurilemmomas occur in cervicofacial region, but intraoral neurilemmomas are extremely rare with only 1% incidence.²

Review of literature provides a variety of terms for neurilemmoma, but only three are still in use: Neurinoma, neurilemmoma, and schwannoma.¹ Neurilemmoma generally arises from cranial and spinal nerve roots or from peripheral nerves but shows predilection for sensory nerves. Almost 25-48% of all schwannomas occur in the head and neck region. The development of this tumor in the mouth is quite uncommon representing only 1% of all head and neck region tumors.² Other common sites include the flexor surface of upper and lower extremities and less often the mediastinum and peritoneum.⁴ On occasion, the tumor can also arise centrally within the bone and may produce bone expansion. Intraosseously, most commonly seen in the posterior mandible and appear as either unilocular or multilocular radiolucencies on radiographs.²

Literature search reveals that neurilemmoma occurs regardless of age and sex, growing gradually, and painlessly. Seldom does it recur, and the malignant transformation is rare.³ Here, we present a rare case occurring in buccal mucosa and discuss the histopathological hallmarks of neurilemmoma.

CASE REPORT

A 54-year-old female patient reported to our institution with a complaint of swelling in the right cheek region since 6 months. History revealed that the onset of swelling was slow, and there was no increase or decrease in the size of the swelling with no association of pain. Intraoral examination revealed a well-defined swelling measuring approximately about 1.5 cm × 1.5 cm. Mucosa over the swelling was normal in color. On palpation, the swelling was firm in consistency, non-tender, non-fluctuant, and non-reducible. The computed tomography scan revealed a well-defined hypoechoic lesion on the right side of the cheek. A provisional diagnosis of benign salivary gland lesion was given. Surgical excision of the lesion was done Gross examination of the surgically excised specimen was white in color, round in shape, firm in consistency measuring about 1X0.8X0.4 cm
On histopathological examination, a well encapsulated lesion consisting of irregularly arranged spindle shaped cells were seen (Figure 2). The majority of cells showed clear cytoplasm with vesicular nuclei, and few cells showed granular cytoplasm (Figure 3). Areas showing Antoni A and Antoni B pattern was very minimal. Few areas showed cystic degeneration. Stroma was interspersed with chronic inflammatory cells, blood capillaries, and nerve bundles. Clear cells were periodic acid-Schiff negative. Immunohistochemical staining with cytokeratin was negative. Further staining with S-100 showed diffuse positivity (Figure 4). Based on the clinical, histopathological and immunohistochemical findings, diagnosis of neurilemmoma was rendered.

**DISCUSSION**

Neurilemmoma, a benign encapsulated nerve sheath neoplasm is composed of Schwann cells which form a thin outline around each extra cranial nerve fiber and wrap larger fibers with an insulating membrane myelin sheath, to enhance nerve conduction. Neurilemmoma arises when proliferating Schwann cells form a tumor mass encompassing motor and sensory peripheral nerves. Wright and Jackson reported 146 cases of neurilemmoma of the oral cavity soft tissue, of which 52% occurred in the tongue, 19.86% in the buccal or vestibular mucosa, 8.9% in the soft palate, and the remainder 19.24% in the gingiva and lip. Verocay in 1910 first described a group of neurogenic tumors, termed as “neurinomas.” In 1935, Stout proposed that these tumors arose from nerve sheath elements and they were termed neurilemmomas. Most reports suggest that the majority of tumors are present between the age of 20 and 40 years and are equally distributed between the both the sexes. There is a predilection for the head, neck, and flexor surfaces of the upper and lower extremities. 25% of the extra cranial neurilemmomas are reported in head and neck region but only 1% in intra-oral region. Clinically, intraoral
neurilemmoma can be of two types-an encapsulated or non-capsulated lesion. Among the intra oral sites affected by neurilemmomas, the tongue is the most common site. Cheek is much rare site of this type of tumor. Literature review by Gallo et al. analyzed 157 cases, wherein 45.2% of the cases involved the tongue and only 13.3% involved the cheek. In our case, the lesion was present in the right buccal mucosa. Neurilemmoma is usually a solitary lesion but can be multiple lesions when associated with neurofibromatosis.

Histologically, neurilemmomas are of five varieties: Common, plexiform, cellular, epithelioid, and ancient schwannomas. Histopathologically, it is an encapsulated tumor demonstrating two cell patterns in varying amounts: Antoni A and Antoni B. Antoni A tissue is characterized by streaming fascicles of spindle-shaped Schwann cells. These cells often form a palisaded arrangement around central acellular, eosinophilic areas known as Verocay bodies. These Verocay bodies consist of reduplicated basement membrane and cytoplasmic processes. Antoni B tissue is less cellular and less organized; the spindle cells are randomly arranged within a loose myxomatous stroma.

Our case due to its anatomical location and histopathological presentation of masquerading features led us to confounding diagnosis of salivary gland lesion like oncocytoma. The histopathological presentation showed large areas of clear cells with vesicular nuclei and few cells with granular cytoplasm sustained the diagnosis of oncocytoma. Support for microscopic diagnosis was obtained by immunostaining with S-100 which showed diffuse positivity demonstrating spindle shaped cells arranged in streaming pattern and presence of large nerve bundle at the periphery of the lesion presenting as classic neurilemmoma.

Immunohistochemistry acts as an adjunct giving the definitive diagnosis of the tumor. All types of neurilemmoma show positivity with neural markers such as S-100 protein and glial fibrillary acidic protein. The pericapsular region of neurilemmoma may also demonstrate CD34-positive cells.

Neurilemmoma is usually well encapsulated, so the treatment of choice is surgical excision under local or general anesthesia depending on the position of the tumor.

The non-encapsulated form requires a margin of normal tissue and careful separation from the involved nerve. As recurrence and malignant transformation of neurilemmoma are exceedingly rare, the prognosis is usually very good. In this case, the mass was well encapsulated and was totally excised. Postsurgical follow-up of the patient showed good prognosis.

REFERENCES