Benign Peripheral Nerve Sheath Tumor of the Tongue

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Peripheral nerve sheath tumors rarely occur in the oral cavity and include neurofibroma, schwannoma, and palisaded encapsulated neuroma. We report a rare case of benign peripheral nerve sheath tumor of the tongue. This tumor was a 0.8 × 0.5 cm sized, firm mass on the left lateral surface of the tongue. Histologically, this mass was composed of well-circumscribed variable sized nodules, which consisted of moderately cellular spindle cells with vague nuclear palisading and a small amount of fibrous tissue. Most of the tumor cells were strongly positive for S-100 protein, but negative for epithelial membrane antigen on immunohistochemistry. No axons were found by immunostaining for neurofilament and Bodian stains. In addition, the surrounding, compressed, fibrous tissue showed rare EMA positive cells. The present case might be a rare case of neurofibroma arising in the tongue, although immunohistochemical and special stains did not support such a diagnosis.

Key Words: Tongue, neurofibroma, diagnosis

INTRODUCTION

Oral peripheral nerve sheath tumors are rare
and include neurofibroma, schwannoma, palisaded encapsulated neuroma (PEN), nerve sheath myxoma, mucosal neuroma associated with multiple endocrine neoplasia III, traumatic neuroma and granular cell tumor. Intraoral schwannoma accounts for 1% of head and neck schwannomas and shows a predilection for the tongue, usually the base region. Schwannoma of the tongue has so far only been described in a few individual cases. PEN represents 0.04 to 0.05% of oral biopsy specimens and most of the lesions are found in the palate region. After the first case of intraoral PEN was described, only a few case reports have been found in the literature. Neurofibroma is an oral manifestation of neurofibromatosis occurring in 4-7% of affected persons, the tongue being the most common site. Although these tumors are distinct entities having characteristic histologic and immunohistochemical features, it has been reported that a spectrum of peripheral nerve sheath tumors existed.

Here we report a case of benign peripheral nerve sheath tumor of the tongue, which was histologically most compatible with neurofibroma, but showed intense S-100 protein positivity on immunohistochemistry in most cells and no axons identified in immunostaining for neurofilament and Bodian stains.

CASE REPORT

A 39 year-old female was admitted due to an asymptomatic tongue mass for 3 months. She had a history of esophageal mass removed by laser operation 5 years previously. Family history was unremarkable. She did not show any other clinical signs or symptoms. Physical examination revealed a 0.8 × 0.5 cm sized, firm mass on the left lateral surface of the tongue. Under the impression of fibroma, excision of the mass was done.

Grossly, a small, whitish gray, firm, solid mass was found in the subepithelial region. Histologically, this mass consisted of four well circumscribed, variable sized nodules (Fig. 1), which were composed of moderately cellular, bland, wavy, spindle cells with a small amount of collagenous tissue (Fig. 2). Vague nuclear palisading...
of the tumor cells was noted, but no verocay body or fascicular arrangement was identified. Several small nerve fascicles were present adjacent to the tumor. Most of the tumor cells were strongly positive for S-100 protein (BioGenex, San Ramon, CA, USA, dilution 1:80) on immunohistochemical stain (Fig. 3). No axons were found in immunostaining for neurofilament (Novacast, Newcastle upon Tyne, UK, dilution 1:50) and Bodian stain. This tumor did not have a distinct capsule, but EMA (DAKO, Carpinteria, CA, USA, dilution 1:100) immunostaining showed rare positive cells in the surrounding compressed fibrous tissue.

**DISCUSSION**

Although peripheral nerve sheath tumors share a common neural origin, they exhibit notable microscopic and pathogenetic heterogeneity. Neuropilidroma is unencapsulated, consisting of a mixture of Schwann cells, perineurial cells, and endoneurial fibroblasts. Schwannoma is an encapsulated tumor composed of Schwann cells arranged in both a cellular palisaded pattern (Antoni type A) with verocay body and a loose paucicellular pattern (Antoni type B). PEN is a well circumscribed nodule, often partly surrounded by a fibrous capsule, with fascicles of spindle shaped cells showing variable and focal nuclear palisading, which consists of proliferation of both Schwann cells and axons. The present case was unencapsulated, histologically uniform and moderately cellular spindle cell tumor. No distinct two patterns, verocay body or fascicular arrangement of tumor cells were identified. These features were more compatible with cellular neuropilidroma than schwannoma or PEN.

The diagnoses of these tumors are usually straightforward, but differential diagnosis may be difficult in some cases and immunohistochemistry and special stain can be useful in diagnosis and
subclassfication of these lesions. S-100 protein is expressed by Schwann cells, but not by perineurial cells or endoneurial fibroblasts, so most of the cells in schwannoma and PEN are intensely stained; an effect which is believed to arise from Schwann cell proliferation. However, neurofibroma showed fewer S-100 protein-positive cells and more variability and might contain scattered EMA-positive cells, in contrast to schwannoma or PEN. In this case, nearly all tumor cells were intensively stained for S-100 protein, thus suggesting a lesion of Schwann cell proliferation, i.e. schwannoma or PEN rather than neurofibroma. However, typical neurofibroma may contain many S-100 protein-positive cells.

Usually, the presence of axon was identified in neurofibroma and PEN but not in schwannoma, thereby distinguishing PEN or neurofibroma from schwannoma. In the present case, axons were not identified by immunostaining for neurofilament and Bodian stains. However, it has been reported that axons could not be demonstrated in some typical PEN due to antibody specificity and extensive microheterogeneity. Moreover, axons might not be identified by immunostaining for neurofilament and Bodian stains in typical neurofibroma. Thus negative staining for neurofilament did not contradict the diagnosis of PEN or neurofibroma.

Therefore, the present case might be a rare case of neurofibroma arising in the tongue, although immunohistochemical and special stains did not support this diagnosis. However, these tumors are completely benign lesions and show no recurrence if completely excised. Therefore, strict differential diagnosis might not be important in the clinical viewpoint.

REFERENCES