An unusual case of neural palatal swelling

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ABSTRACT

Neural tumors in the oral cavity occur both in the soft tissues and in the jaw bones. They occur as painless, smooth surface swelling in the soft tissues of the mouth, exhibiting a slow rate of growth and mild expansion of the cortical plates. Here, we report a rare case of a cellular variant of schwannoma in a young Indian female patient who presented with an asymptomatic nodule in the mid-palatine raphe region of the hard palate, mimicking a fibroma, thus creating a dilemma in the diagnosis.

Keywords: Nerve sheath tumor, palatal swelling, schwannoma, Schwann cells, spindle cell tumor

Introduction

Swellings in the palate may result from a variety of etiological factors and can originate from the structures within the palate or beyond it, which may simulate each other in various forms making the diagnosis challenging for the clinicians.¹ The most commonly encountered swellings in the anterior palate are the soft tissue tumors, odontogenic abscess, cysts, and tumors. In this case, since the clinical and radiographic investigations ruled them out, some of the neural tumors were considered, out of which the diagnosis of cellular schwannoma was confirmed after immunohistochemistry. Although the occurrence of cellular schwannomas is rare in the oral cavity, they may present as a benign tumor in the majority of the cases.

Case Report

A 30-year-old female patient presented with a 8 months history of a swelling located in the midline of the hard palate (Figure 1). The swelling was persistent and firm with no increase in the size over a period of 3 months. There was no history of trauma, bleeding, or discomfort from the lesion. Family history and review of systems were non-contributory.

On examination, no changes were seen extra-orally. Head and neck examination demonstrated no palpable cervical or submandibular lymph nodes. Intra-orally, a well-defined solitary nodular swelling of size 1 cm × 1 cm was present in the region of the mid-palatine raphe. The overlying mucosa was slightly erythematous at the periphery. On palpation, the swelling was nontender, firm and attached to the underlying mucosa. The surface of the swelling was smooth and was neither compressible nor reducible (Figure 1).

Based on the above clinical findings, a provisional diagnosis of a benign mesenchymal non-odontogenic tumor was considered. A fine needle aspiration cytology revealed hemorrhage with few squamous cells suggesting no evidence of inflammation or malignancy. A maxillary occlusal radiograph and digital volumetric tomography did not reveal any significant finding in relation to maxillary anterior teeth.

The differential diagnosis considered was benign soft tissue tumors like fibroma, neural tumors such as neurofibroma and traumatic neuroma. A benign minor salivary gland tumor such as a pleomorphic adenoma was considered on account of its site, their slow growths, and absence of neural symptoms. The possibility of oral focal mucinosis though rare was also included.
An excisional biopsy was performed under general anesthesia, and it was subsequently followed by a histopathological examination to conclude the provisional diagnosis. The histopathological examination revealed a highly cellular well-circumscribed encapsulated spindle cell tumor, which is separated from the overlying surface epithelium by fibrous stroma (Figure 2). Sheets of spindle cells with vesicular nuclei, intermingled with ovoid cells are arranged in the form of fascicles (Figure 3). At the periphery of the lesion, peripheral nerve bundles are seen. These features are suggestive of cellular schwannoma. Immunohistochemical evaluation revealed intense nuclear staining for S-100. More than 50% of cells were positive and confirmed the diagnosis (Figure 4). The patient was followed up after 10 days with satisfactory healing and no fresh complaints.

**Discussion**

Schwannoma or neurilemmoma is a benign neoplasm that is derived from a proliferation of Schwann cells of the neurilemma or nerve sheath. As the lesion grows, the nerve is pushed aside and does not become enmeshed within the tumor. This lesion is an encapsulated submucosal mass that presents typically as an asymptomatic lump in patients of any age. The tongue is the favored location, although lesions have been described throughout the mouth. It has been reported that only 1% of these involve the oral cavity. It commonly affects cutaneous tissues of the head and neck region and is classified into four histopathological variants: Conventional, cellular, plexiform, and melanotic.²

Head and neck schwannomas may occur at any age but are most commonly found during the second and third decades of life with no gender predilection, though exceptional cases (usually the cellular variant) may recur locally. This variant of schwannoma has been mistaken for sarcoma (at least in the past) in up to 30% of cases.³

Cellular schwannomas commonly affect soft tissues such as the retroperitoneum and posterior mediastinum and also bone but are extremely rare in the oral region. Koizumi et al. reported a case of cellular schwannoma in the buccal vestibule of a 34-year-old Japanese man, while Oh et al. in 2008 reported a case of cellular schwannoma in the lower lip region of a 27-year-old woman, which presented as a soft papule.⁴,⁵ Similarly, Ogütcen-Toller et al. reported cases of

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**Figure 1:** Clinical image shows a nodular swelling in the mid-palatine raphe of the hard palate

**Figure 2:** Histopathological image shows well-encapsulated spindle cell tumor along with normal marginal tissue seen (H and E, ×4)

**Figure 3:** Histopathological image shows sheets of spindle cells with vesicular nuclei intermingled with ovoid cells arranged in the form of fascicles (H and E, ×40)

**Figure 4:** Immunohistochemical image shows intense nuclear staining for S-100. More than 50% of cells were positive (IHC stain, ×40)
cellular schwannoma involving the mandible. In this case, the 30-year-old woman presented with a solitary nodular swelling in the mid-palatine raphe region of the hard palate which was firm and asymptomatic, mimicking a fibroma thus creating a dilemma in the diagnosis.

The differential diagnosis of traumatic neuroma was ruled out as there was no history of trauma in the region. To rule out neurofibroma, immunohistochemical staining for mast cells was done with toluidine blue which was negative. Palatal pleomorphic adenoma presents clinically as a painless, slow-growing mass found on posterior lateral aspect while the site, in this case, was the mid-palatine raphe. The possibility of oral focal mucinosis was eventually ruled out on the basis of histopathology as spindle cells were observed, favoring the diagnosis of cellular schwannoma which was further confirmed with immunohistochemical evaluation.

Unlike classical schwannoma, cellular schwannoma discloses a marked increase in cellularity, encompassing fascicles of spindle cells, which can occasionally be associated with herringbone or storiform pattern. First described in 1969 by Harkin and Reed Sand later by Woodruff et al., the cellular schwannoma has become a well-recognized variant of the schwannoma because of its cellularity, mitotic activity and the occasional presence of bone destruction have been misdiagnosed as malignant in more than one-fourth of cases.

Oh et al. in his case report revealed that the tumor is histopathologically characterized by the presence of compact spindle cells arranged into fascicles, variable nuclear hyperchromasia and pleomorphism, lack of Verocay bodies and the typical predominance of Antoni A areas. Similarly, in our case, histopathological examination revealed highly cellular encapsulated spindle cell tumor (Antoni A) with numerous fascicles along with peripheral nerve bundles near the capsule, these features concluded the diagnosis of cellular schwannoma and the same was confirmed after immunohistochemical staining. Immunohistochemical evaluation revealed intense nuclear staining for S-100 in which more than 50% of cells were positive.

As cellular schwannomas are of a benign nature, the treatment of choice is complete surgical excision with particular attention to the preservation of the originating nerve, if encountered. In this case, the mass was well-encapsulated and could be totally excised under general anesthesia. The patient was followed up after 1 year with no signs of recurrence.

**Conclusion**

This case highlights the importance of considering the various benign spindle cell neoplasms in the differential diagnosis of palatal swellings. Even though the occurrence of cellular schwannomas in the oral cavity is very rare, it is important for the clinicians to emphasize the possibility of neural lesions and confirm their final diagnosis with histopathological and immunohistochemical evaluation for appropriate treatment and management of the patient.

**References**