Case Report

Lingual schwannoma in an adolescent girl- A diagnostic challenge

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Abstract

Shwannoma, a benign neurogenic neoplasm consisting of schwann cells is a slow growing solitarily found pathology. We present a case of shwannoma in a 14 year old girl and its identifying features which can be a diagnostic challenge owing to its similarity with many lesions. The patient visited our center with a nodular growth on her tongue. Transoral excision was performed and pathologic examination confirmed the diagnosis of schwannoma.

Keywords: Neurilemmoma, schwannoma, tongue neoplasm

Introduction

Shwannoma, shwanoma, or otherwise called neurilemmoma is a benign neurogenic neoplasm consisting of schwann cells. It is a slow growing, encapsulated, well demarcated, solitarily found rare entity. Among reported cases, about 30% are found in head and neck region with just 1% being reported in oral cavity. A total of 152 cases of schwannoma of tongue has been reported in literature over past 60 years.[1] Lingual shwannomas generally are seen in the third decade of life with a 33% incidence and display slightly more predilection in women. They most often present as a soft painless mass (69%). Origin is mostly from sensory nerves and can affect all the cranial nerves, barring optic nerve and olfactory nerve. In tongue, differentiation between glossopharyngeal or lingual nerve origin is difficult considering their proximity. Location and size of the tumorous lesions determine the presence and intensity of the symptoms. Treatment goal should almost always be complete excision.

Case Report

A 14-year-old girl was referred to oral diagnostic department of Riyadh Elm University by a private practitioner to investigate a soft protruding mass on her tongue. Patient had no history of burning sensation, pain or paresthesia in the region. Nodule had been present for 2 years. Size of the nodule increased slowly over 2 years which was noticed by patient at the age of 12 years as a small growth. Blood investigations were normal. Only complaint of the patient was dysphagia. There was no history of bleeding from the lesion. Patient never noticed any change in color over the period of 2 years. She did not report any other growth in her mouth or other areas of the body.

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Clinical examination revealed a globular mass on dorsal surface of tongue measuring 2.0 cm × 3.0 cm × 3.0 cm [Figure 1]. Lesion was soft in consistency. Patient's only concern was mild discomfort at the site of swelling. There was no cervical lymphadenopathy. Growth in the transition area between the oral section of tongue and the base of the tongue was smooth and fluctuant. Surrounding lymph nodes were not palpable. Proposed treatment was transoral excision under local anesthesia.

Excision

Transoral excision was performed as an outpatient procedure under local anesthesia. One cartridge of Infiltrative anesthesia (lidocaine 2%) was administered around the lesion. We excised the lesion with a circular incision [Figure 2] around the margin of the defect with a 2-cm margin below the surface of the tongue. Lesion was excised in total including a 3 mm margin [Figures 3 and 4].

Postoperative period was uneventful with good functional results [Figure 5]. Histopathology showed a circumscribed submucosal nodule with spindle cells and thin wavy nuclei with stratified squamous epithelium showing both Antoni type A with Verocay bodies and Antoni type B areas suggesting a schwannoma. Schwannomas have a dual pattern of tumor arrangement with a well-defined capsule [Figure 6]. One pattern (Antoni type A), shows Schwann cells arranged in palisades, interlacing fascicles, and organoid pattern resulting in Verocay bodies. Another pattern (Antoni type B), shows the Schwann cells loosely packed and embedded in an oedematous stroma. There has been no recurrence of the lesion 2 years following the excision.

Discussion

Schwannoma is a slow-growing benign tumor of the nerve sheath. It is usually a single, circumscribed, firm, painless lesion of variable size.[1] Causative factors of schwannomas are not known. About 30% of all schwannomas occur in the head and neck,[2] with the parapharyngeal space being the most common location.[3] Schwannomas are rare in oral cavity, but if it occurs, is usually seen in the tongue.[4,5]

Most schwannomas present as solitary, painless mass. When multiple schwannomas occur, however, they can be associated with neurofibromatosis. Schwannoma occurs more commonly in females (slight preponderance) than males, and known to occur at any age, majority falling in the 6th to 7th decades.[6]

Histologically, schwannomas display several features. Most schwannomas are encapsulated, and beneath this capsule, two main patterns are intermingled yet sharply defined from each other. First pattern is referred to as Antoni type A, which consists of closely packed Schwann cell in bundles or rows with palisading, elongated nuclei. Free bands of amorphous substance between rows of nuclei constitute the Verocay bodies. The second pattern is known as Antoni type B and is composed of loosely arranged Schwann cells set in a meshwork of reticulum fibers and microcysts.[7]

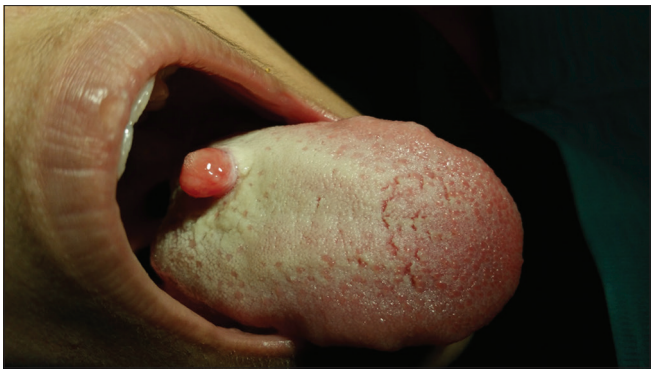

Figure 1: Lesion *in situ*

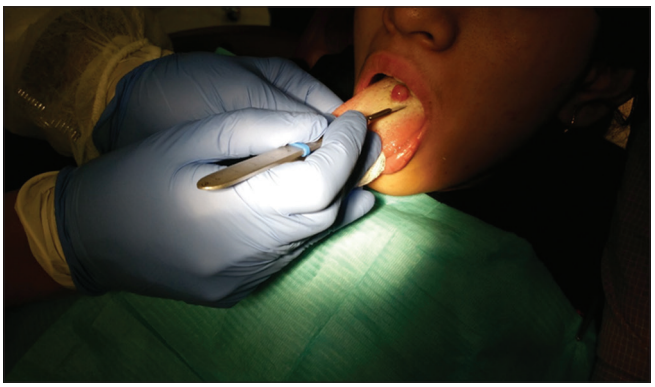

Figure 2: Incision placement

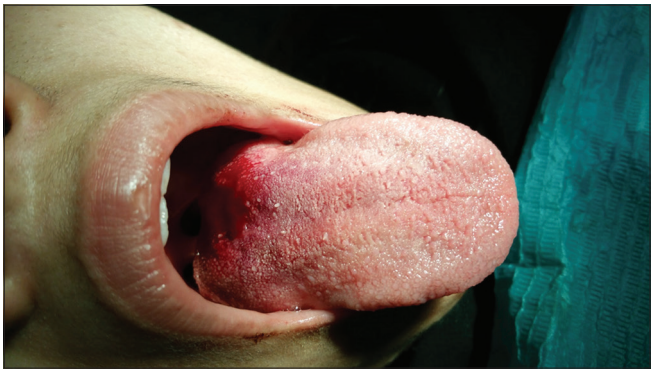

Figure 3: Immediate post-operative excision site

Differential diagnosis of schwannoma includes neurofibromas, granular cell tumors, irritation fibromas, leiomyomas, rhabdomyomas, hemangiomas, lymphangiomas, lipomas, pyogenic granulomas, and benign salivary gland tumors.[8] Since shwannoma mimics hemangiomas, primary care of the lesion and quick diagnosis is essential to rule out such alarming lesions. Histological differential diagnosis include other neural tumors such as neuroma, neurofibroma, or muscular or fibroblastic origin tumors. An excisional biopsy with histopathological evaluation is necessary for confirmation. Differentiation between neurofibroma and shwannoma is necessary because a solitude neurofibroma can be a manifestation of neurofibromatosis that may have malignant transformation.[9] The absence of epithelial-myoepithelial elements, mucoid matrix, smooth muscles, and reticulin

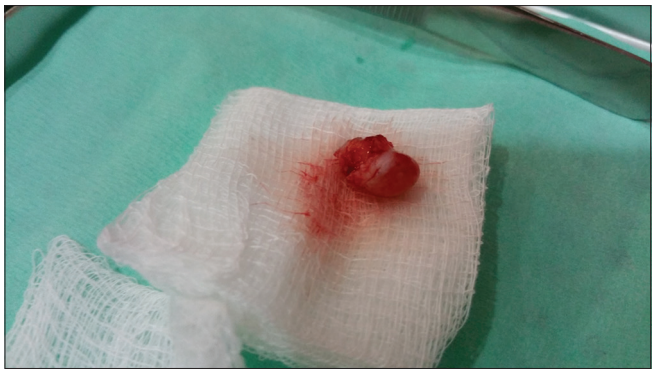

Figure 4: Excised tumor

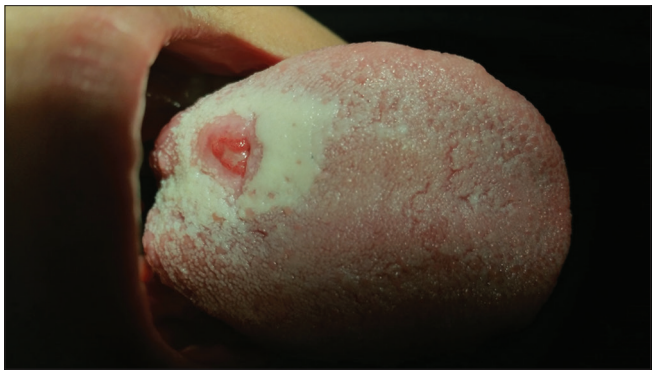

Figure 5: One day post-operative site

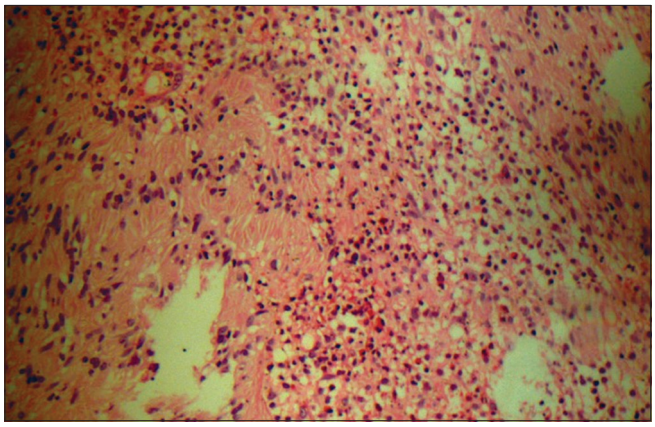

Figure 6: Histopathologic impression

distinguishes shwannoma from similar other histologic entities. Complete excision of schwannomas have excellent prognosis. Tongue tumors being accessible to FNA, can be rapidly examined cytologically with regard to their benign or malignant nature.[10] Focal changes of anaplastic cells, numerous mitoses, increased cellularity, and invasiveness suggest malignant transformation. In the case presented here, excision has been complete and no sign of recurrence seen two years post-surgical procedure. Transoral resection with care to preserve the nerve remains the standard protocol.[11] There are reports of carbon dioxide laser excision being used to treat base of tongue schwannomas.[12] In this case, tumor removal was complete without mucosal damage to avoid tongue dysfunction and recurrence.

Conclusion

Shwannoma of the tongue is considerably rare. Transoral excision of this tumor precludes recurrence, avoids morbidity of tongue function and remains the standard approach for treatment of majority of these tumors. Potential for malignant transformation of this tongue tumor is 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.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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