SCHWANNOMA OF THE BUCCAL MUCOSA

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ABSTRACT

Schwannoma is a rare benign nerve sheath tumor originating from Schwann cells of the peripheral nerves. Intra-oral development of this tumor accounts for only 1% of all the head and neck tumors. A preoperative diagnosis of schwannoma is not possible due to its rare occurrence. In this paper we present a case of a 5 year old girl with a schwannoma of the lower right buccal mucosa, diagnosed on the basis of intra-operative clinical findings and histopathological examination and treated via an intra-oral surgical approach.

Key Words: Schwannoma, Schwann cells, Benign Nerve Sheath Tumor, Buccal mucosa.

INTRODUCTION

The schwannoma of buccal mucosa is a benign neurogenic tumor rarely reported in the literature. Almost 25-45% of all schwannomas occur in the head and neck region but the development of this tumor intra-orally is rare which accounts for only 1% of all head and neck region tumors. The most common intraoral sites in order of decreasing frequency is the tongue, palate, floor of mouth, buccal mucosa, lips and the jaws.

The definitive diagnosis of schwannoma preoperatively is difficult in soft tissue since clinically it shows striking resemblance with fibroma, lipoma and neurofibroma. A final diagnosis can only be established with the help of histopathological examination. Surgical excision is the modality of choice for treatment of schwannoma. This article presents a rare case of schwannoma of the buccal mucosa in a very young child and a review of the literature.

CASE REPORT

A 5 year old girl reported to the Out Patient Department of Oral and Maxillofacial Surgery, Abbasi Shaheed Hospital, Karachi with the chief complain of a swelling on her lower right side of face since one year. The swelling was painless and asymptomatic and had gradually increased in size over a period of one year. She had no history of associated facial trauma.

Fig 1: Extra Oral picture showing swelling on lower right side of face.

Fig 2: Intra-oral picture showing the vestibular incision in right mandibular buccal sulcus.
or infection and her past dental, medical, family and social history were not significant.

On extra-oral clinical examination a 3.5 x 3.5 cm firm swelling was present on the lower right side of her face near the chin. The swelling was non-tender, lobulated with irregular smooth margins and was attached to the skin. There was no paresthesia of the overlying as well as the surrounding skin. (Fig 1) On intra-oral examination a soft tissue bulge was palpable in right mandibular buccal sulcus with no odontogenic cause.

Surgical intervention was carried out under general anesthesia. The lesion was approached via an intra-oral vestibular incision at the lower right mandibular buccal sulcus (Fig 2). Intra-operatively the lesion was found to be an encapsulated, pink nodular growth extending from canine to 1st molar on right side of mandible (Fig 3). The lesion was completely excised. It had a broad base of about 3.5 x 1 cm in size attached with an elongated cylindrical mass of 2.5 x 2.5 x 1 cm (Fig 4). There was no evidence of attachment to the neurovascular bundle. Primary closure was done.

The patient was re-evaluated on the next day after the surgery, post-operative findings were unremarkable and no neuronal deficit was evident. Microscopically, the tissue consisted of a well-circumscribed neoplastic lesion arranged in (Antoni B) hypo- and (Antoni A) hypercellular areas associated with scattered Verocay bodies.

### TABLE 1: SUMMARY OF CASE REPORTS FOR SCHWANNOMA OF BUCCAL MUCOSA INCLUDING THE PRESENT CASE

<table>
<thead>
<tr>
<th>No.</th>
<th>Authors</th>
<th>Age, Sex</th>
<th>Location</th>
<th>Duration</th>
<th>Follow Up</th>
</tr>
</thead>
<tbody>
<tr>
<td>1.</td>
<td>Yasuyuki et al11, 2002</td>
<td>34, M</td>
<td>Left Buccal Mucosa</td>
<td>Unknown</td>
<td>No recurrence</td>
</tr>
<tr>
<td>2.</td>
<td>Subhashraj et al9, 2009</td>
<td>18, M</td>
<td>Left Cheek lower Vestible</td>
<td>8 months</td>
<td>No recurrence</td>
</tr>
<tr>
<td>3.</td>
<td>Kim et al9, 2011</td>
<td>66, F</td>
<td>Left Buccal Mucosa</td>
<td>13 years</td>
<td>No recurrence</td>
</tr>
<tr>
<td>4.</td>
<td>Pravin et al5, 2013</td>
<td>52, M</td>
<td>Left Cheek</td>
<td>6 months</td>
<td>No recurrence</td>
</tr>
<tr>
<td>5.</td>
<td>Suchitra et al12, 2014</td>
<td>45, M</td>
<td>Left Cheek</td>
<td>1 year</td>
<td>No recurrence</td>
</tr>
<tr>
<td>6.</td>
<td>Arwade et al7, 2014</td>
<td>40, M</td>
<td>Left Mandibular Vestibule</td>
<td>1 year</td>
<td>No recurrence</td>
</tr>
<tr>
<td>7.</td>
<td>Anureet et al13, 2016</td>
<td>49, M</td>
<td>Mucosa of Cheek</td>
<td>15 years</td>
<td>No recurrence</td>
</tr>
<tr>
<td>8.</td>
<td>Hala et al, 2017 (this present case)</td>
<td>5, F</td>
<td>Right lower Buccal Mucosa</td>
<td>1 year</td>
<td>No recurrence</td>
</tr>
</tbody>
</table>
hyper-cellular areas. The spindle shaped cells had a moderate amount of cytoplasm and hyperchromatic variable nucleoli. Scattered prominent Verocay bodies were also identified (Fig 5). Therefore a classical diagnosis of schwannoma was confirmed. The patient is under regular follow-up and there has been no recurrence after three months.

**DISCUSSION**

Schwannomas, also referred to as neurinomas or neurilemmomas are benign, solitary, slow growing, encapsulated nerve sheath tumor composed of Schwann cells. Schwannoma is a tumor of unknown etiology.

During the fourth week of embryological development these Schwann cells are derived from a specialized population of ectomesenchymal cells of the neural crest. These cells form a covering around each extracranial nerve fiber and envelope larger fibers with an insulating membrane, the myelin sheath. The main purpose of this sheath is to intensify nerve conduction. The myelination of nerve inside the brain and spinal cord is carried out by oligodendrocytes but as the nerve exits, myelination is done by Schwann cells. If during the proliferation the Schwann cells aggregate into a tumorous mass around motor and sensory peripheral nerves, Schwannoma appears.

It has no gender propensity and can occur at any age. However, it is more common between the third and fourth decades of life. William et al findings showed that in 83% of the cases studied by them the schwannomas presented in males, while for Lucas there was a greater predilection for females, and for Hatziotis and Asprides; Enzinger and Weiss there was an equal distribution between both sexes. In 50% of the cases a direct relation with the involved nerve can be seen.

Gallo et al reported on 157 cases, where 45.2% of the cases involved the tongue and 13.3% involved the cheek. Gupta et al on 136 cases of schwannoma in the head and neck that consisted of 60 cases in the neck, ten cases in the parotid gland, nine cases in the cheek, eight cases in the tongue, and eight cases in the pharynx. Kun et al reported in their study that 18 out of 49 cases were in the neck and 11 in the tongue. Wright and Jackson studied 146 cases of schwannoma of the oral cavity soft tissue. Out of which, 52% involved the tongue, 19.86% the buccal or vestibular mucosa, 8.9% the soft palate, and the remainder 19.24% were in the gingivae and lip.

In the present case schwannoma was located in the right mandibular vestibule arising from the buccal mucosa presenting as an ovoid, slow growing, asymptomatic swelling with irregular margins, all features that point towards some more commonly seen benign soft tissue lesions such as mucocele, fibroma, lipoma, low grade salivary gland neoplasm or neurofibroma. Hence, after clinical examination schwannoma was not predicted in the provisional diagnosis as it is a rare finding especially in the buccal mucosa of vestibule.

Excisional biopsy was planned as the diagnostic as well as therapeutic treatment modality in this case.

The histopathological findings of this case were comparable to the ones reported earlier in literature, consisting of a well-circumscribed neoplastic lesion arranged in hypo- and hyper-cellular areas namely Antoni type A and type B. The Antoni type A tissue consist of Schwann cells that are closely packed, with elongated, palisaded nuclei. Scattered prominent Verocay bodies which are composed of bands of amorphous substance were present between the rows of nuclei. Under electron microscope, Verocay bodies show cytoplasmic projections composed of small amount of collagen and basal laminar material. The Antoni B tissue has lower cell count and is not well organized, the spindle shaped cells are widely separated and dispersed with a network of delicate reticulated fibers. The vasculature is generally not prominent, changes like inflammation, fibrosis, and nuclear atypia are seen in some old tumors.

In this case, histopathology of the entire specimen shows a majority of Antoni B cells with prominent Verocay bodies that confirms the classical diagnosis of Schwannoma. An additional confirmatory test for Schwannoma according to literature survey, is immunohistochemical analysis for presence of S100 protein. According to Chrysomali et al the tumor cells with Antoni A have higher intensity scores compared to Antoni B tumor pattern.

Although all tumors of neural origin have S100 protein positive on immunohistochemical examination but it can prove to be helpful in differentiation of lesion. Chrysomali et al reported, in schwannoma and encapsulated neuroma there is an intense positive reaction to S100. Intensive reaction to CD57 is seen in traumatic neuroma, whereas in schwannoma capsular epithelial membrane antigen (EMA) and CD34 stainings are seen.

Passador-Santos et al studied S100 protein, EMA, laminin, fibronectin, and collagens I and II in neural benign neoplasms. According to the study, schwannoma cells were positive for S100 regardless of the growth pattern. EMA was seen only in the capsule staining perineurial cells and laminin stained basement membrane around tumor cells. Fibronectin and collagens I and III were expressed, especially in the capsule. In neurofibroma, the cells expressed S100 whereas EMA was expressed only in a few scattered cells. Laminin and fibronectin showed a diffuse positivity. Collagens I and III showed a fibrilar arrangement with diffuse positivity.
Schwannoma of the buccal mucosa

The gold standard for treatment of Schwannoma is the complete surgical excision. According to Asaumi et al, diagnostic investigations such as ultrasonography, CT scan and MRI are useful, in order to estimate margins of tumors, composition of tumor, and to determine the relation with surrounding structures. There is no recurrence and transformation into a malignancy is rare. Prognosis is good.

When the diagnosis of a Schwannoma is established it is of utmost importance to examine the patient for development of a tumor elsewhere in the body, although in most cases it’s not seen. Schwannoma should be differentiated from neurofibroma, since a solitary neurofibroma can be associated with neurofibromatosis.

After a review of the literature few of the related case reports are summarized in Table 1. All except for the present case occurred in adults, there was a similar clinical presentation of schwannoma in these cases i.e. asymptomatic mass, increased gradually over a period of months, misdiagnosed preoperatively, treated with surgical excision and final diagnosis of schwannoma established after histopathological examination and immunohistochemical analysis.

CONCLUSION

Intra-oral Schwannoma represents a rare pathology of the head and neck region, not clinically differentiable from most benign soft tissue lesions in this area. Definitive diagnosis is only possible after a histopathological analysis and in certain cases immunohistochemical assay for S100 protein. Complete surgical excision is the suggested treatment modality for schwannoma.

REFERENCES


CONTRIBUTIONS BY AUTHORS

1 Syeda Hala Raza Jafri: Article writer, Conception, design and interpretation of data.
2 Sufyan Ahmed: Operating Surgeon, Final approval of the manuscript.