Cellular Schwannoma of Buccal Mucosa: A Rare Histopathological Variant

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ABSTRACT

Introduction: Schwannoma (Neurilemmoma) is a benign neoplasm that develop from schwann cells in the peripheral nerve sheath. It commonly occurs as an encapsulated, slow-growing and generally solitary lesion. Cellular schwannoma is a rare histopathological variant of schwannoma.

Case Presentation: Here, we discuss a case of 44-year-old female patient who reported with the chief complaint of swelling in the left upper back cheek region for the past 2 years. Histopathological and immunohistochemical analysis confirmed the diagnosis as cellular schwannoma.

Management and prognosis: Surgical excision of the lesion was performed and no recurrence was reported after 1 year of follow up.

Conclusion: Cellular schwannoma a rare intraoral benign tumor, needs to be differentiated from other malignant tumor with a careful approach for a prompt diagnosis and proper management of the lesion.

Key Words: Cheek, Neoplasm, Neurilemmoma, Schwann cell

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Introduction

Schwannoma is a benign, encapsulated, slow-growing, and generally solitary tumor that arise along the course of any nerve in the body associated with schwann cells. It is usually a painless lesion with smooth surface, and its clinical symptomatology depend on the nerve of origin. Approximately 25-45% of all schwannomas are seen in the head and neck region but less than 1% occur in the oral cavity. The high cellularity and co-existence of several growth patterns such as fascicular, storiform, and herringbone patterns contributes to the difficulty in recognition as schwannoma. Among the histopathological variants, cellular schwannoma seems to be a rare histopathological variant of schwannoma.1 In the present article, we report a rare case of cellular schwannoma, in a 44 -year- old female patient occurring in buccal mucosa which is not associated with any syndrome. The voluminous and painless appearance of the lesion as well as associated microscopic features with hypercellularity and pleomorphism may lead to a false impression of a malignant tumor. Hence it is necessary to have a thorough examination to confirm the benign nature of the tumor thereby avoiding unnecessary treatment.

Case Report

A 44 year old female patient reported to the outpatient department with a chief complaint of painless swelling in the left upper jaw for the past two years. Past medical and family history was non-contributory. Extraoral examination revealed facial asymmetry corresponding to the swelling on the left side of the face. The swelling was well-circumscribed extending superiorly

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from the left zygomatic arch to inferiorly till 2cms away from the corner of the lip. Horizontally the lesion was extending from ala of the nose till external acoustic meatus. No lymph nodes were palpable. On intraoral examination, the lesion showed a well-circumscribed mass on the left buccal mucosa in relation to 26, 27, and 28 measuring 2 x2 cm (Fig. 1). It was firm in consistency and non-tender on palpation. No pus discharge was evident. Radiographic examination revealed radiolucency in relation

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to 26, 27 and 28, MRI and CT findings showed large lobulated hyperintense lesion measuring 4x4x5 cm in the left side of inferior temporal fossa, lesion caused smooth indentation over the lateral wall of maxilla and was abutting the 3rd molar on left side (Fig. 2). Based on the clinical findings, the differential diagnosis varied from

Fig. 1: Clinical photograph showing swelling in left buccal mucosa in relation to 26, 27, and 28 measuring 2 x2 cm

benign lipomatous tumors to benign tumors of peripheral nerves and malignancy.

Surgical excision of the lesion was done under general anaesthesia. The lesion was enucleated in toto and sent for histopathological examination. Macroscopically, the gross specimen consisted of a soft tissue mass which was brownish-black in color, irregular in shape, and soft in consistency. The cut surface of the tumor appears homogenous, solid, as myxoid and white in color (Fig. 3). Histopathological examination revealed compact fascicular proliferation of schwann cells with occasional palisaded nuclear arrangement resembling Antoni A type with the lack of verocay bodies (Figs. 4a & 4b). The mitotic activity was comparably low. The entire lesion was covered by a well-defined capsule. Immunohistochemistry showed positive expression for S100 (Fig. 5) and negative for Ki-67 and a confirmatory diagnosis of 'Cellular Schwannoma' was made. The postoperative course was uneventful with no signs of recurrence.

DISCUSSION

The term 'Schwannoma' had been attributed in the past either for neurofibroma or neurilemmoma. But the present concept is that the first one originates from perineural cells and the latter one from schwann cells and the term schwannoma can be interchangeably used with neurilemmoma. 2 It is usually seen in the second and fifth decade but is also reported in early childhood, the case presented also occurred in fourth decade. They have equal distribution

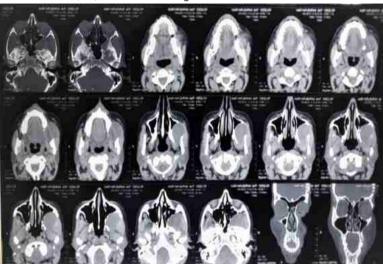
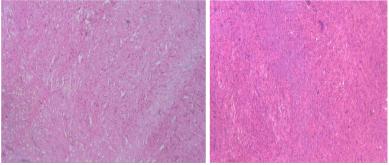


Fig. 2: Pre-operative MRI - large lobulated hyper intense lesion measuring 4x Fig. 3: The gross specimen 4x5 cm present in the inferior temporal fossa of left side





Figs. 4a & 4b: Long fascicles of schwann cells without Antoni B areas or verocay bodies(H & E staining; x100)

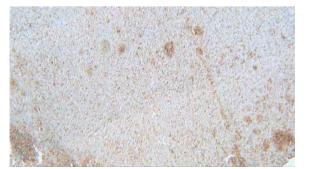


Fig. 5: Positive for S-100 (IHC; x100)



in male and female with slight female preponderance as in the present case. Cohen et al., in 2009 reported that almost 25%-45% of all schwannomas occur in the head and neck region. Only 1% are seen intraorally of which tongue is the most common site but the present case reported in buccal mucosa. Clinically it appears as a slow-growing, solitary, non-indurated, well-circumscribed lesion with a smooth surface.³

The histopathological variants of schwannoma are ancient pseudoglandular schwannoma, schwannoma, cellular schwannoma, epithelioid schwannoma, and melanotic schwannoma. Woodruff et al in 1981 reported a series of 14 cases of cellular schwannomma that were histologically characterized by increased cellularity, nuclear pleomorphism, hyperchromatism, lack of verocay bodies, and frequently higher mitotic activity.⁴⁻⁶ The present case showed all these features described by Woodruff et al except higher mitotic activity. In contrast, our case showed low mitotic activity. Sudhir Bhalerao et al in 2012 reported that the mitotic activity usually does not exceed four per ten high power fields as was in our case. Immunohistochemically, the tumor cells are diffusely immune-reactive to S-100 protein as in most reported cases of cellular schwannoma. Diffuse and strong immunoreactivity to S-100 protein in the present case confirms the neural origin of the tumor¹.

The treatment of choice is excision. Hribernik et al in 1992 reported that encapsulated form is enucleated easily, whereas the non-encapsulated lesion requires normal tissue margins to avoid relapse.⁷ If the nerve of origin is visualized, an attempt should be made to separate carefully in order to preserve the function, although may not be possible at times. The prognosis is very good since it does not usually recur and malignant transformation is rare. In the present case, the lesion is encapsulated and postsurgical healing was normal despite the large size of the lesion and no recurrence was reported after 1 year of follow up. Kun et al in 1993 described six cases in the maxillofacial region, two of these had malignant transformation, but in the case reported no malignant transformation was seen.^{8,9}

CONCLUSION

Schwannomas are slow-growing; painless masses that arise

commonly in the head and neck region and the oral cavity. While dealing with an intraoral, well-defined soft tissue mucosal lesion, the possibility of schwannoma should be kept in mind and distinguished from other benign as well as malignant lesions that can develop in the oral cavity. Histopathological and immunohistochemical analysis are essential for a definite diagnosis. The prognosis for the lesion is favourable and local resection is mostly curative.

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