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Case Report

Pathological Diversity in Schwannomas of the Orofacial Region

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Abstract

Schwannoma is a slow-growing, encapsulated benign tumor of the neuroectodermal origin arising from the perineural Schwann cells. This study aims to elucidate the clinicoradiographical and histopathological features of orofacial schwannomas through a case series of seven cases. The patients' aged ranged from 13 to 45 years, with a male predilection in the ratio of 5:2. One intraosseous case presented as a radiolucent lesion. All the cases exhibited Antoni A and Antoni B type of microscopic patterns in varying amounts. One case of ancient schwannoma showed degenerative features. The tumor cells showed diffuse positive immunohistochemical reaction for S-100 protein. Our study suggests that intraosseous schwannoma should be considered in the differential diagnosis of the intraosseous jaw lesions. Histopathologically, it is important to recognize the findings of ancient schwannoma and to avoid misdiagnosing it as a malignant lesion.

Keywords: *Ancient schwannoma, Antoni A and Antoni B areas, intraosseous schwannoma, S-100 protein, Verocay bodies*

Introduction

Schwannoma or neurilemmoma is a slow-growing, encapsulated benign tumor of neuroectodermal origin arising from the perineural Schwann cells.[1,2] It was first named neurinoma by Verocay in 1910. Term schwannoma was introduced by Masson (1932). Later, the term neurilemmoma was coined by Stout (1935).[3] Approximately 25%–45% of cases are seen in the head and neck region, of which 0.5%–1% are found within the oral cavity. Intraorally, tongue is the most common site involved. It usually presents as an asymptomatic, solitary submucosal mass in the oral cavity.[1,4] Intraosseous presentation is rare (less than 1%), with mandible being more commonly involved.[5]

Histologically, schwannoma is usually an encapsulated tumor, consisting of mixture of two cellular patterns: Antoni type A and Antoni type B.[6] Antoni type A tissue is characterized by streaming fascicles of palisaded spindle-shaped Schwann cells with twisted nuclei around central acellular eosinophilic areas known as Verocay bodies. Antoni type B areas are characterized by less cellular and less organized spindle or oval cells within a loose myxoid hypocellular matrix.[3,6] Conventional schwannomas that exhibit hyperchromatic, atypical, and pleomorphic nuclei along with areas of hemorrhage and hemosiderin are diagnosed as ancient schwannoma. Ancient schwannoma is an unusual variant of schwannoma.[7]

Material and methods

Archival data of seven cases of schwannoma were retrieved from the files of the Department of Oral Pathology, Maulana Azad Institute of Dental Sciences, New Delhi. 3-µm thick sections of paraffin-embedded tissues were stained with hematoxylin and eosin (H and E) and reviewed.

Immunohistochemistry (S-100, vimentin, desmin, smooth muscle actin [SMA], and cytokeratin) was performed by conventional standard technique with streptavidin–biotin immunoperoxidase method to confirm diagnosis in instances where H and E stain was insufficient for a confirmatory diagnosis.

In the present case series of seven cases, all lesions were soft in consistency, present in different regions of head and neck including tongue, pterygomandibular

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raphe, mandibular anterior tooth region, palate, preauricular region, and maxillary vestibule with one intraosseous lesion involving anterior maxilla [Figure 1]. All the lesions were slow growing (range of 3 months to 9 years) with a mean size of 2.5 cm × 2.2 cm. 71.4% of te patients were male with age ranging from 13 to 45 years.

Macroscopically, the cut sections showed homogenous gray-white areas [Figure 2]. The characteristic microscopic features of schwannoma were seen in all the cases. 71.4% of cases were encapsulated. The mixture of Antoni A and Antoni B areas was found in all the cases. 85.7% of cases showed foci of myxoid changes and 57.1% of the cases showed microcyst formation. Mild-to-moderate chronic inflammatory cell infiltrate was found in all the cases [Figure 3a and b]. One case (Case 1) showed degenerative features including nuclear atypia, pleomorphism, and hyalinization and was thus diagnosed as ancient schwannoma [Figure 3c and d]. To confirm the diagnosis, immunohistochemistry was done by using S-100, vimentin, cytokeratin, desmin, and SMA [Figure 3e and f]. The clinical findings are summarized in Table 1 and histopathological findings are summarized in Table 2.

Discussion

Schwannomas are solitary tumors and usually originate from the proliferation of Schwann cells in the perineurium of the peripheral, cranial, or autonomic nerves, which usually result in displacement and compression of the adjacent nerves.[1] The pathogenetic mechanism responsible for tumor is loss of function of merlin, the protein encoded by the neurofibromatosis type 2 (NF2) gene. Loss of function of merlin, either by direct genetic change involving the NF2 gene on chromosome 22 or secondarily to merlin inactivation, results in downstreaming of its signaling pathways which lead to formation of tumor.[2]

Schwannomas occur most frequently in middle-aged individuals (25–55 years).[3,4] In our case series, the age ranged from 13 to 45 years with M: F ratio of 5:2 showing male predominance. Similar studies were reported by Williams *et al*., where males were affected more frequently.[8] Study by Lucas found a female predominance, while other studies found no gender predilection.[4,9] Based on their location, two types of schwannomas are described: peripheral (extraosseous) and central (intraosseous).[10,11] Many authors found that tongue is the most frequent site of occurrence.[4,12,13] In the present case series, all the lesions were present on different regions of head and neck.

Clinically, these soft tissue tumors may be mistaken for other benign lesions such as peripheral ossifying fibroma, traumatic fibroma, and pleomorphic adenoma. In the current case series, the tumor was present as soft tissue submucosal nodule on different sites in the head and neck region, with one case of central schwannoma. Less than 1% of lesions are intraosseous and predominantly involve the mandible. It is considered that there are three mechanisms by which schwannomas may involve bone: (a) a tumor may arise centrally within bone, (b) a tumor may arise within the nutrient canal and cause canal enlargement, or (c) a soft tissue or periosteal tumor may cause secondary erosion and penetration into bone.[14,15] Radiographically, intraosseous schwannomas are difficult to differentiate from other bone lesions such as fibrous dysplasia, neurofibroma, central giant cell lesion, or periapical lesion.[14] Maxillary schwannomas are extremely rare. To the best of our knowledge, only 13 cases of maxillary schwannomas have been reported till date. The central schwannoma in our case series involved the anterior maxilla (Case 3).

Histopathologically, schwannomas are unilocular encapsulated masses. In the current case series, all the cases showed features of conventional schwannoma consisting of two cellular patterns: Antoni type A and Antoni type B.[2,16,17]

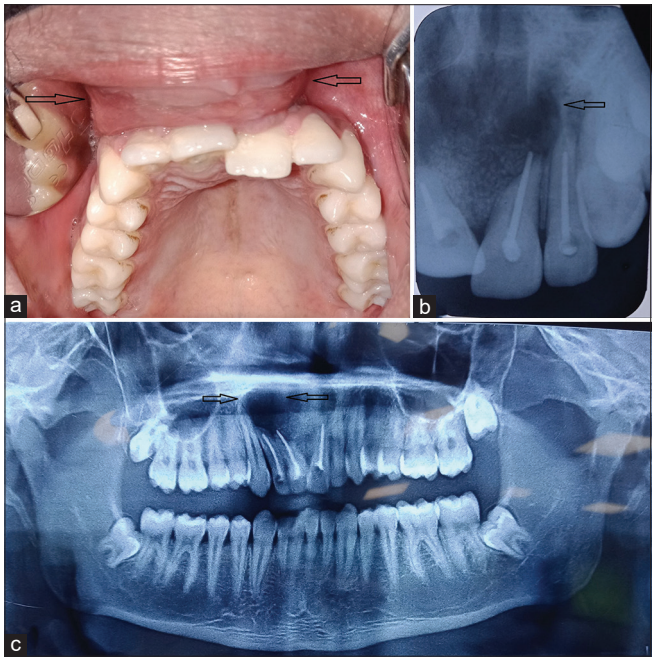

Figure 1: Intraosseous schwannoma. (a) Solitary swelling of approximately 4 cm × 3 cm was observed in the anterior maxilla in relation to #11, 12, and 21 teeth. On palpation, it was nontender and bony hard in consistency with no fluctuation or softening in any part of the swelling. (b) Periapical radiograph mimicking a presentation of periapical pathology. (c) Orthopantomograph revealed ill-defined radiolucent lesion with interrupted corticated border in relation to #11, 12, and 21 teeth (Case 3)

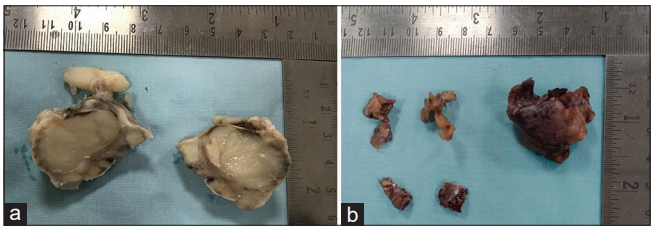

Figure 2: (a and b) Macroscopically tissue was globoid shaped and tan in color. It consisted of a capsule and homogeneously firm in texture (Case 3 and Case 4)

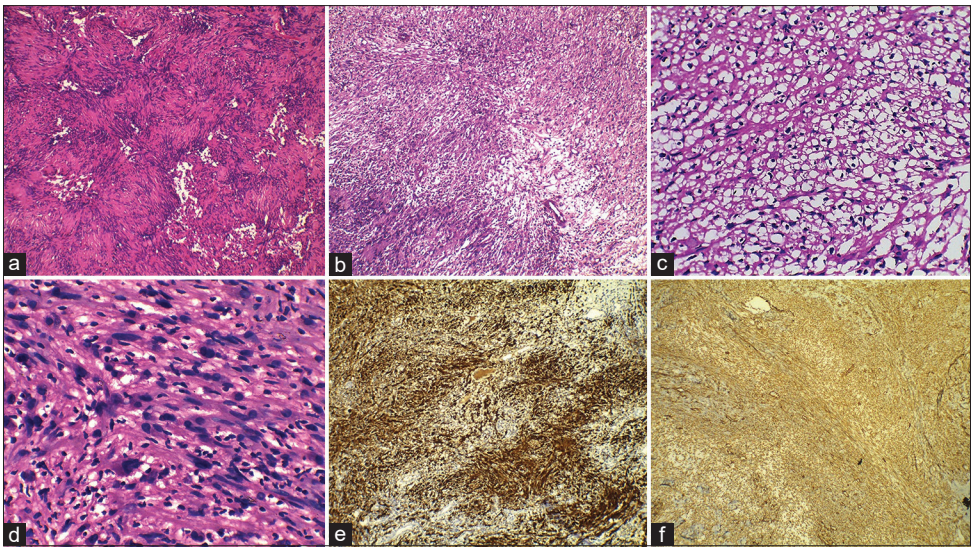

Figure 3: (a) Photomicrograph showing Antoni type A areas composed of spindle-shaped tumor cells with palisaded nuclei surrounding the amorphous eosinophilic central Verocay bodies. (b) Antoni type B areas composed of less cellular and less organized loosely fibrous background. (c) Ancient schwannoma consisting of degenerative features – microcyst formation and (d) cellular pleomorphism and nuclear atypia. (e) S-100–positive expression. (f) Positive expression of vimentin

Table 1: Summary of clinical features of schwannomas

Case number	Age	Sex	Site	Duration	Size (cm)	Provisional diagnosis
1	20	Male	Tongue	3 years	2×1	Traumatic fibroma
2	30	Female	Right pterygomandibular raphe	8-9 years	5×5	Traumatic fibroma
3	18	Male	Anterior maxilla	1.5 years	3×2	Nasolabial cyst
4	13	Male	Mandibular anterior tooth region	3 months	3×4	Peripheral ossifying fibroma
5	45	Male	Right preauricular region	3 months	2×2	Lipoma
6	18	Male	Soft palate	15 days	1×1	Fibroma
7	30	Female	Right maxillary vestibule	7 years	1×1.5	Spindle cell tumor

Table 2: Summary of histopathological features of schwannomas

Features	Case number						
	Case 1	Case 2	Case 3	Case 4	Case 5	Case 6	Case 7
Capsule	Thin	Thick	Thin	Thin	Thin	-	-
Antoni A	+	+	+	+	++	++	+
Antoni B	++	++	++	+	+	+	+
Verocay bodies	+	+	+	+	++	++	+
Myxoid changes	+	+	+	+	+	+	-
Degenerative changes	++	-	-	-	-	-	-
Microcyst formation	++	+	+	+	-	-	-
Inflammatory cells	++	+	+	++	+	++	+
IHC	S-100 vimentin	S-100	S-100	S-100	ND	S-100	S-100 vimentin

+: Less present; ++: More present; -: Absent. ND-Not done; IHC-Immunohistochemistry

Ancient schwannoma is an unusual variant of schwannoma. It was first described in the thorax by Ackerman and Taylor in 1951.[7] It is a rare benign encapsulated long-standing tumor. Histologically, it consists of degenerative changes along with presence of Antoni A and Antoni B cellular areas. Degenerative features are represented by areas of hemorrhage, hemosiderin deposits, inflammation, fibrosis, hyalinization, and nuclear atypia.[2,7,16,17] It is believed that long history of the lesion could be the cause of the transformation to an "ancient" variant.[18] Out of current case series, one case (Case 1) showed features of ancient schwannoma. The duration of the lesion was 3 years. Histopathologically, it showed cellular atypia, nuclear hyperchromasia, and pleomorphism with some areas showing interstitial hyalinization.

Immunohistochemically, the cases showing S-100 (highly reactive) and vimentin (weakly reactive) positivity were

diagnosis as schwannoma. S-100 is an acidic protein which usually stains the neural crest derivatives. It is consistently expressed in schwannomas as majority of the cells of schwannomas have Schwann cell antigenic phenotype.[8]

In conclusion, we recommend that intraosseous schwannoma should be considered in the differential diagnosis of intraosseous lesions of the head and neck region. As ancient schwannoma shows degenerative features, it is important to recognize the histopathological findings to reach the correct diagnosis. The treatment option for schwannoma is surgical removal, and recurrence after local excision is rare.

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Conflicts of interest

There are no conflicts of interest.

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