CASE REPORT

Ancient neurilemmoma: A rare oral tumor

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ABSTRACT
Neurilemmomas are benign tumors of neural origin composed of Schwann cell proliferation in characteristic patterns. Ancient neurilemmomas are usually longstanding growths that exhibit degenerative features that could be mistaken for malignancy. They are extremely rare in the oral cavity and present in older individuals of long duration. The authors report a case of ancient neurilemmoma in a young patient with short duration of growth. This unique case presented with remarkable histopathological features with respect to vascularity and atypia associated with degenerative change. It is essential to not mistake these features as malignant transformation so as to avoid radical procedures.
Key words: Ancient schwannoma, neurilemmoma, neural tumor

INTRODUCTION

Oral schwannomas are uncommon benign tumors, composed of Schwann cell proliferation. The microscopic picture is characteristic, described as a spindle cell lesion with Antoni A and Antoni B arrangements.[1-5] Some lesions named "ancient," present with degenerative features. Oral ancient schwannomas are exceedingly rare.[6-10] The management and clinical behavior is the same as classic schwannoma. Recurrence is rare. We report a case of ancient schwannoma highlighting the histopathology. The unique feature in our report is the short duration of the tumor in a young patient, which might raise suspicion of malignancy.

CASE REPORT

A 22-year-old male patient reported to the dental facility with a growth in the right back tooth region of his mouth, He had noticed the start of the growth 6 months ago. The lesion was stated to be painless and slow growing. On intraoral examination there was a nodular soft tissue mass in the mucobuccal fold region, in the right posterior maxillary quadrant above the molars. It was about 2 × 2 cm in size, smooth, sessile, non-tender and firm on palpation, with slight bleeding on manipulation. A provisional diagnosis of pyogenic granuloma was made [Figure 1a]. The patient consented to surgical excision and the surgery was performed after general clinical and routine laboratory investigations. He is being followed-up regularly for the past 3 months and progress has been uneventful.

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The cut-section of the excised tissue revealed a solid lesion with hemorrhagic areas [Figure 1b]. On histopathological examination, the lesion presented as a circumscribed cellular mass [Figure 2]. The cells were mostly spindle shaped with oval/blunt elongated nuclei. They were arranged in two patterns. One pattern was a palisaded arrangement of cells encircling eosinophilic structures (suggestive of Verocay bodies) [Figure 3]; and the other pattern was an irregularly arranged configuration of spindle cells, sometimes in a loose myxoid background [Figure 4a and b]. These patterns appeared consistent with descriptions of Antoni A and Antoni B patterns observed in schwannomas. There was significant vascularity throughout the lesion, with thick-walled blood vessels surrounded by hyalinization and inflammatory cells [Figure 5a and b]. Extravasated red blood cells (RBCs), hemosiderin deposition and siderophages were occasionally encountered [Figure 6]. In some areas, the spindle cells revealed nuclear pleomorphism and increased nuclear cytoplasmic ratio [Figure 7a and b]. Mitotic activity was insignificant, with mitotic index (MI) <1 per high power field (HPF). No abnormal or bizarre mitotic figures were observed.

Taking into account the above features of schwannoma, along with extensive hemorrhage, hemosiderin deposition and cellular atypia, a histopathological diagnosis of ancient schwannoma was given.

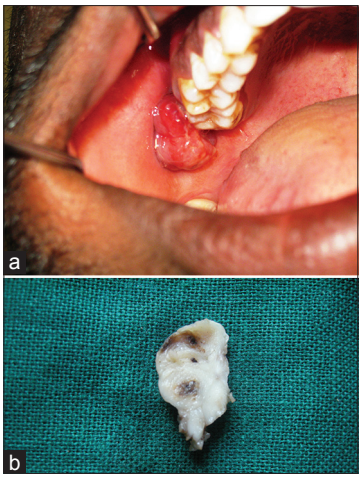

Figure 1: Clinical photograph of the oral lesion (a). Cut section of the excised specimen showing the solid and hemorrhagic areas (b)

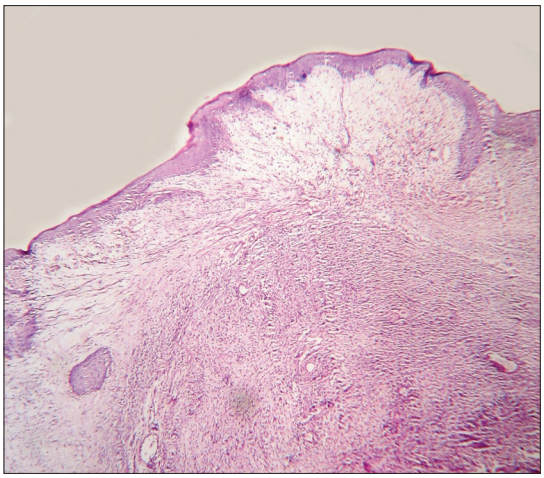

Figure 2: Photomicrograph of the circumscribed lesion (H&E stain, ×40)

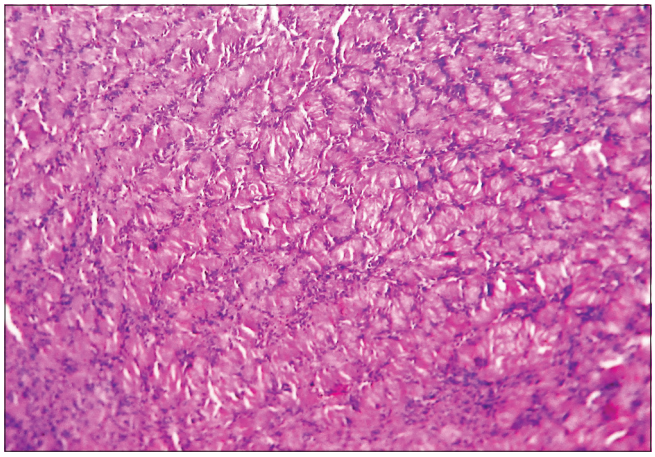

Figure 3: Photomicrographs of the Antoni A areas. (H&E stain, ×100)

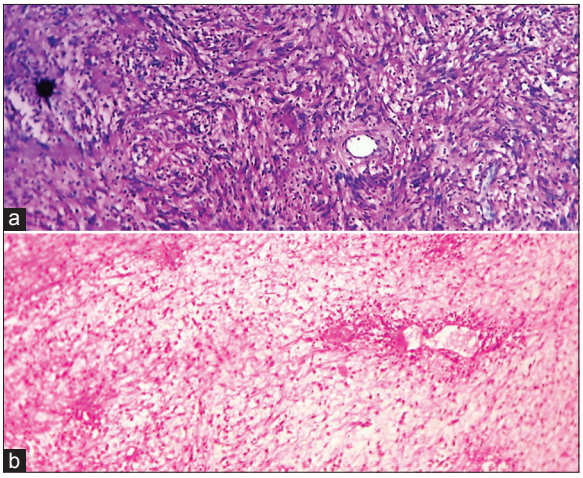

Figure 4 (a and b): Photomicrographs of the Antoni B areas. (H&E stain, ×100)

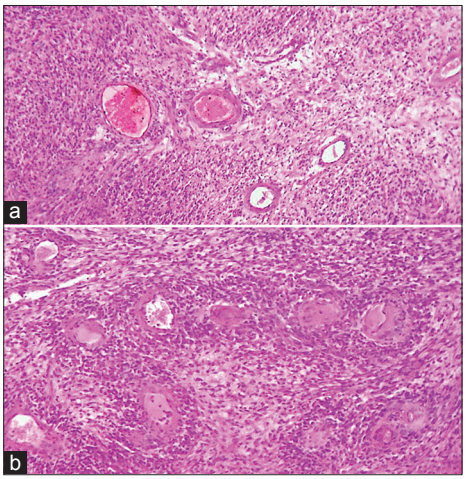

Figure 5: Photomicrographs of the vascularity (a) and prominent hyalinization and inflammatory infiltrate (b) (H&E stain, ×100)

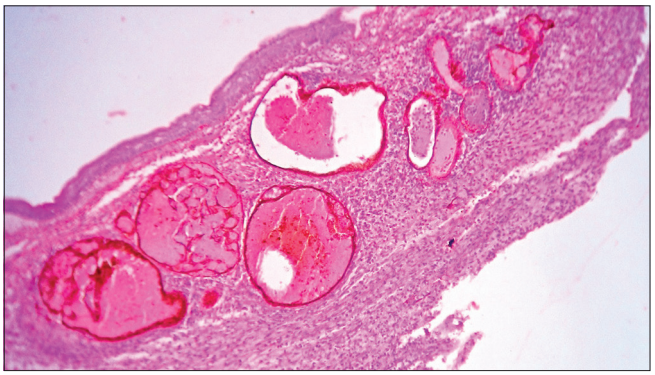

Figure 6: Photomicrographs of the large vessels with hemorrhage and hemosiderin. (H&E stain, ×40)

DISCUSSION

The schwannoma is a benign tumor of the neural Schwann cells that can potentially occur in any part of the body. It is said to develop from the neural sheaths of cranial (except optic and olfactory), peripheral motor, sensory and sympathetic nerves.[1] Up to 25% may be associated with Von Recklinghausen's Neurofibromatosis II (NFM II). In the head and neck region (25-45% cases) it can involve the facial soft tissues, sinuses, nasal and oral cavities, parotid,

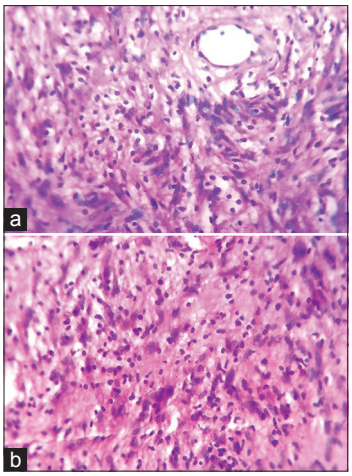

Figure 7 (a and b): Photomicrographs of the cellular atypia. (H&E stain, ×400)

pharynx and cervical nerve trunks.[2] Oral cavity tumors are quite uncommon, comprising up to 1% of the total.[3,4] The ancient variety is extremely rare in the oral cavity, with only 16 cases reported so far.[5-10] The oral schwannoma is seen mostly in the tongue but can occur in the labial and buccal mucosa, palate, vestibules, etc., It usually presents as a slowly enlarging nodule.[3,4]

Histopathologically, the tumor is biphasic, consisting of Antoni A and Antoni B patterns. Antoni A pattern consists of fascicles of palisaded bipolar Schwann cells streaming around acellular eosinophilic areas. These cells usually have oval or blunt elongated nuclei. The acellular zones assuming organoid shape are termed Verocay bodies. The Antoni B pattern is less cellular, with randomly arranged cells in a loose myxoid stroma. Associated nerve and neurites may be seen peripherally, but are not a part of the tumor. Vascularity is also a feature of schwannomas.[1-5]

Some workers classify schwannomas into five types: common type, plexiform, cellular, epithelioid and ancient. The plexiform schwannoma has a multinodular growth pattern, which is cellular and prone to recurrence. The cellular schwannoma has a predominant Antoni A pattern with atypical features and increased mitosis. Both of these types are benign and should not be mistaken for sarcomas.[1,11]

In lesions where hemorrhage, hemosiderin deposition and chronic inflammatory changes are observed with pleomorphic cells, the term ancient schwannoma is given. The ancient schwannoma was described first by Ackerman and Taylor in 1951. The oral ancient schwannoma was first reported by Eversoll and Howell in 1971.[6] Only few cases have been reported in the oral cavity. In all these reports the patients were in their 4th decade of life or older. The mean age of occurrence was 43 years and the usual size of the tumor ranged from 0.9 to 5.5 cm. The average duration of presentation was 10 years, but in some cases 2-5 month duration had been noted.[6-9] The microscopic alterations reported were cystic degeneration, prominent myxoid areas, bizarre spindle cells and occasional mitoses. The atypical changes are due to degeneration and should not be misconstrued as malignancy. Large hyalinized thick-walled blood vessels, hemorrhagic foci, lipid laden and hemosiderin-laden histiocytes are not uncommon. The cells themselves are spindle shaped, with eosinophilic cytoplasm and tapered nuclei; some of which are pleomorphic and hyperchromatic. Necrosis can be seen. Immunohistochemistry may be performed to confirm the diagnosis.[11,12]

Malignant peripheral nerve sheath tumor (MPNST) is a malignancy of neural tissue, whose histological features resemble the fibrosarcoma. Microscopic evidence of invasion is required for diagnosis. Frequent local recurrence and distant metastasis are part of its typical biological behavior.[13-15] In the present case, the diagnosis of ancient schwannoma was made due to the presence of cellular atypia, vascular pattern, hemorrhage, and hemosiderin accumulation; notwithstanding the duration of only 6 months. Factors apart from chronicity may be responsible for the atypical changes observed in this case.

Schwannomas stain strongly with S-100 (97%), Leu-7 (57%) and myelin basic protein (MBP) (44%), confirming their neural origin. However, neurofilament (NF) is usually negative due to absence of neurites. This feature can distinguish schwannomas from the neurofibroma, where nerve fibers are a part of the tumor.[6] CD 56 and calretinin are usually positive in schwannomas, while CD 34 and Factor XIIIa are positive in neurofibromas. These markers can be included in the panel in doubtful cases. In case of MPNSTs, S-100 is usually positive, except in the perineural variant, where epithelial membrane antigen (EMA) showed strong reaction in perineural cells. Markers of proliferation like increased MIB-1 labeling, p53 overexpression and loss of retinoblastoma (RB) protein expression have been found in MPNST case series.[13-15]

Electron microscopy shows bipolar cells with long cytoplasmic processes, entangled with extracellular matrix like collagen and basement membrane material characteristic of Schwann cells. The stroma sometimes consists of long-spaced collagen bundles with a 130 nm banding pattern, which are termed Luse bodies.[12,16]

CONCLUSION

Schwannomas, including the ancient variety, are treated by surgical excision. Recurrence is rare. Ancient schwannomas may be mistaken for malignant change; therefore deeper knowledge of this variety is crucial to avoid unnecessary surgery. Most cases of recurrence are due to inadequate excision, and some are associated with NFM-I1 or Multiple endocrine neoplasia (MEN) type III.[1,16]

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