American Journal of Case Reports

e-ISSN 1941-5923
© Am J Case Rep, 2022; 23: e938335
DOI: 10.12659/AJCR.938335

Received: 2022.09.07
Accepted: 2022.11.21
Available online: 2022.12.01
Published: 2022.12.23

Ancient Schwannoma: Case Report of an Unusual Entity in an Unusual Oral Location

Authors' Contribution:
Study Design A
Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
Literature Search F
Funds Collection G

ABDEFG **Faraj Alotaiby**

Department of Maxillofacial Surgery and Diagnostic Sciences, College of Dentistry, Qassim University, Qassim, Saudi Arabia

Corresponding Author: Faraj Alotaiby, e-mail: f.alotaiby@qu.edu.sa
Financial support: None declared
Conflict of interest: None declared

Patient: Male, 21-year-old
Final Diagnosis: Ancient schwannoma
Symptoms: Facial swelling
Medication: —
Clinical Procedure: —
Specialty: Dentistry • Pathology

Objective: Rare disease

Background: Schwannoma is an uncommon benign tumor that develops from Schwann cells in the peripheral nervous system; 25-50% of such tumors occur in the head and neck. A rare variety of schwannoma, known as an ancient schwannoma, exhibits calcification, cystic degeneration, hemorrhage, myxoid stroma, pleomorphism, and nuclear hyperchromatism. Such characteristics may contribute to this tumor being incorrectly diagnosed as malignant. Almost all reported intra-oral ancient schwannomas behave innocuously.

Case Report: A 20-year-old man presented to the oral surgery clinic with a right facial swelling that began a few months before. CT and MRI were performed and showed a well-defined cystic lesion anterior to the right masseter muscle, located in the subcutaneous area and measuring 28×24×20 mm. No extension to the surrounding tissues nor any other similar lesions were seen. Microscopic examination revealed an encapsulated nodular mass with cystic cavities, hemorrhage, and variably distributed myxoid and hyalinized stroma. The tumor demonstrated 2 distinct cell distributions: hypercellular areas (Antoni A) and myxoid hypocellular areas (Antoni B). Focal areas in high-power magnification showed atypical cells with hyperchromatic and pleomorphic nuclei. The tumor cells showed diffuse positivity for S-100 in IHC. The diagnosis of ancient schwannoma was made. At 1-year follow-up, there was no evidence of nerve injury or recurrence.

Conclusions: One more case of oral ancient schwannoma is added to the literature. This is the first reported in Saudi Arabia. It is prudent to be familiar with the degenerative and cytologic alterations of ancient schwannoma, as this may limit the likelihood of misdiagnosis and unnecessary treatment of this benign tumor, with an excellent prognosis.

Keywords: Schwann Cells • Schwannomatosis

Full-text PDF: https://www.amjcaserep.com/abstract/index/idArt/938335

Indexed in: [PMC] [PubMed] [Emerging Sources Citation Index (ESCI)] [Web of Science by Clarivate]

Background

Schwannoma, also known as neurilemmoma, is an infrequent benign tumor that develops from Schwann cells in the peripheral nerves. Although they are uncommon, 25-50% of such tumors occur in the head and neck. They typically manifest in the oral cavity as asymptomatic nodules. Both genders are equally affected, with the greatest incident reported between the ages of 20 and 40 years [1,2].

A rare variety of schwannoma known as an ancient schwannoma exhibits calcification, cystic degeneration, hemorrhage, myxoid stroma, pleomorphism, and nuclear hyperchromatism. Such characteristics may contribute to this tumor being incorrectly diagnosed as malignant. Almost all intra-oral ancient schwannomas behave innocuously [3].

A systematic review investigated the published articles related to oral ancient schwannoma, revealing only 27 reported cases from 1950 to 2016. The age range was 11-82 years, with average age 40 years. The male: female ratio was 1: 2. The most common reported oral location was the floor of the mouth. Other reported sites include the tongue, jawbone, palate, submandibular area, and labial and buccal mucosa [4].

Most cases are asymptomatic; however, paresthesia and pain were documented in 4 cases. Dysphagia and voice change was reported in 2 cases, located in the submandibular area and the tongue. Of 26 case reports, 17 were reported in Asia, including India, Iran, Korea, Japan, Taiwan, and Israel. No cases were reported in Saudi Arabia [4].

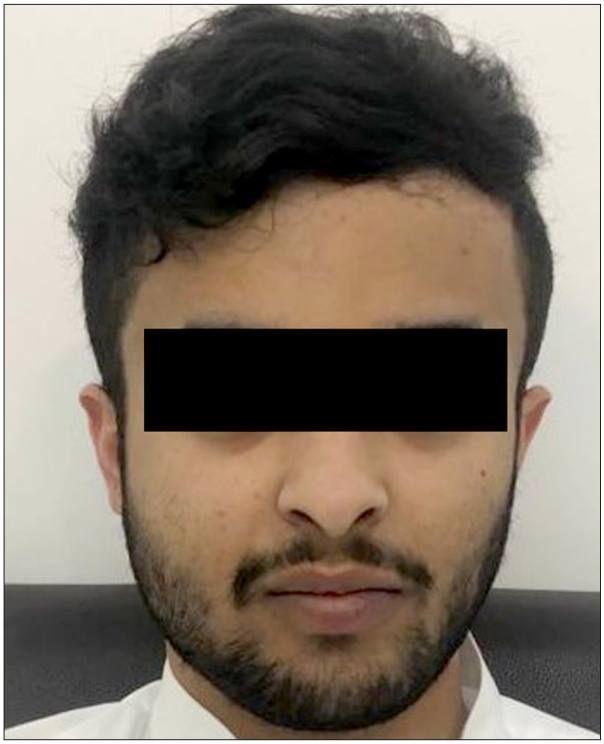

Figure 1. Facial asymmetry due to swelling in the right cheek.

The present case report presents the clinical, radiographic and histologic features of a newly reported case of ancient schwannoma in the buccal mucosa of 21-year-old man with a 1-year follow-up in Qassim, Kingdom of Saudi Arabia.

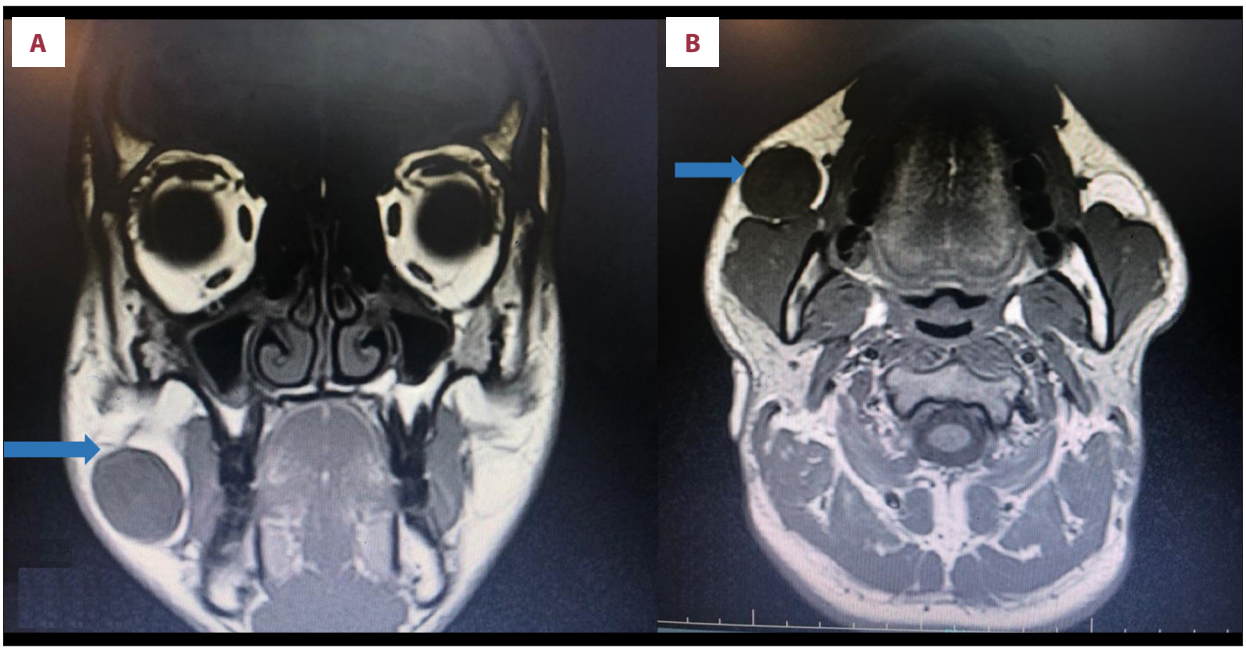

Figure 2. (**A, B**) Coronal and axial MRI show a well-defined lesion anterior to the masseter muscle (arrows).

Indexed in: [PMC] [PubMed] [Emerging Sources Citation Index (ESCI)] [Web of Science by Clarivate]

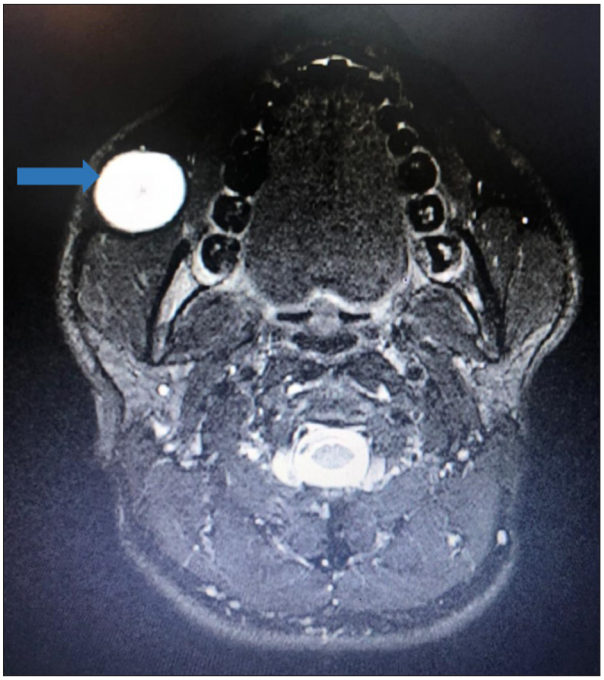

Figure 3. MRI shows a cystic lesion anterior to the right masseter muscle, measuring 28×24×20 mm (arrow). Hypointense in T1 and hyperintense in T2 with internal mini-cystic areas.

Case Report

A 20-year-old man attended the oral surgery clinic, Doctor Tooth Dental Center, Bukairiah, Qassim, Saudi Arabia, with a right facial swelling that began a few months before. The patient had no known medical illnesses or significant family history. This swelling caused a conspicuous asymmetry of the face (**Figure 1**). No pain was reported. An extra-oral examination revealed a mass formation in the soft tissue of the right cheek, close to the parotid duct. The lesion was firm and could be demarcated by palpation. Lymph nodes were impalpable. An intra-oral examination showed smooth-surfaced swelling that was slightly movable upon manipulation on the right-side buccal mucosa, with almost 3 cm in diameter.

Radiographic studies, including ultrasonic, CT, and MRI, were performed and showed a well-defined cystic lesion seen anterior to the right masseter muscle located in the subcutaneous area, measuring 28×24×20 mm. No extension to the surrounding tissues nor any other similar lesions were seen. Hyperintense T2 and low-intense T1 denoted the cystic nature, with minute internal mini-cystic areas (**Figures 2-4**).

The differential diagnosis includes benign neoplasms, either of salivary origin, like pleomorphic adenoma or cystadenoma, or soft-tissue neoplasms like myofibroma, solitary fibrous tumor, or any other mesenchymal neoplasms.

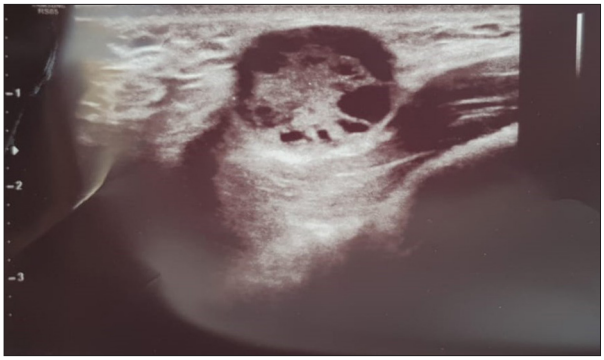

Figure 4. Ultrasonography shows a hyperechoic heterogeneous round mass without any internal flow.

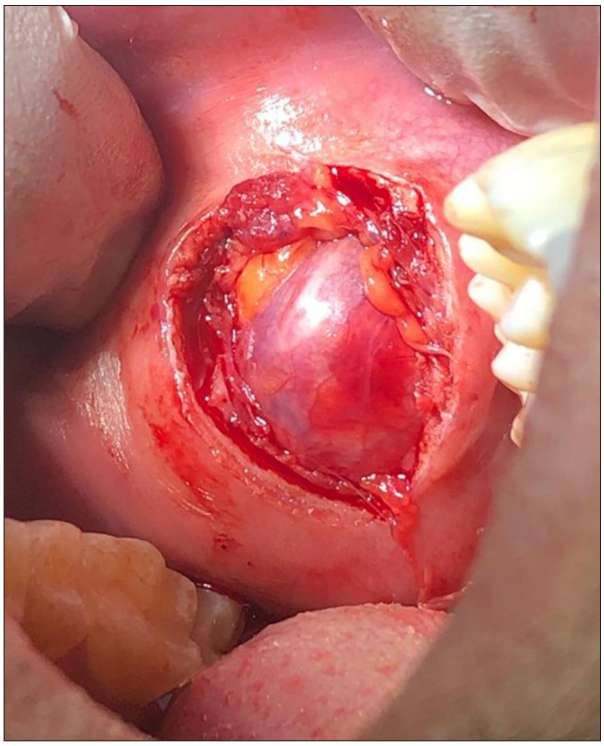

Figure 5. Intraoperative view of the lesion.

The lesion was uneventfully surgically resected under local anesthesia. The lesion was located just beneath the oral buccal mucosa, was well capsulated, and not attached to the surrounding tissue; therefore, it was easily separated from the surrounding tissue. The eradicated lesional tissue was then submitted for microscopic examination (**Figure 5**).

Upon gross examination, the specimen appeared as a single nodular tan piece measuring 2.8 cm in the greatest dimension. Multiple small cystic cavities were noted upon sectioning (**Figure 6**).

Microscopic examination revealed an encapsulated nodular mass with cystic cavities, hemorrhage, and variably distributed

Indexed in: [PMC] [PubMed] [Emerging Sources Citation Index (ESCI)] [Web of Science by Clarivate]

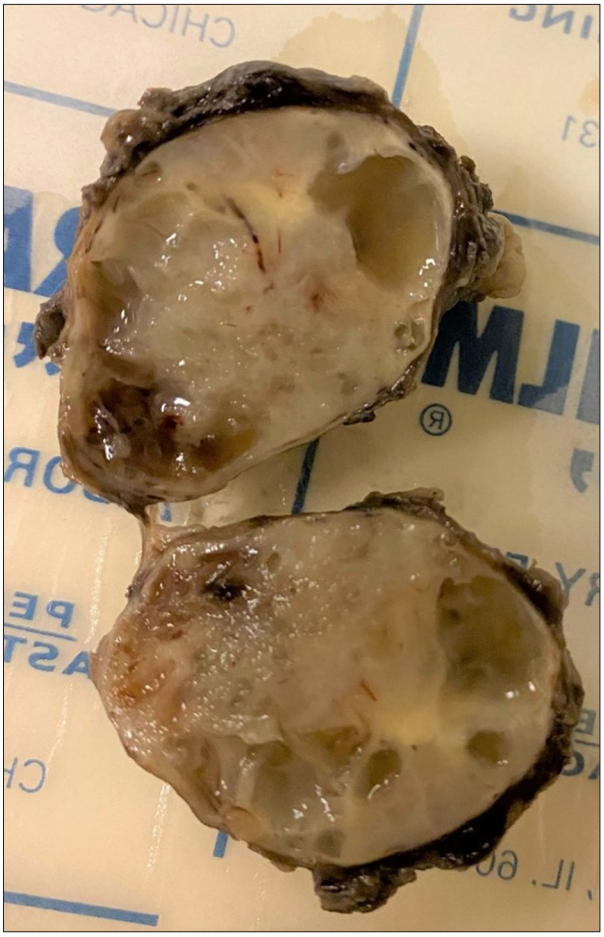

Figure 6. Gross section of the lesion showing surrounding capsule and cystic cavities with hemorrhage within the lesion.

cells in myxoid and hyalinized stroma. The tumor demonstrates 2 distinct cell distribution: (1) hypercellular areas with nuclear palisading and Verocay bodies (Antoni A), and (2) myxoid hypocellular areas (Antoni B). Large, haphazardly spaced blood vessels and hemorrhage were prominent, particularly in hypocellular myxoid areas. The tumor cells were fine, elongated, and wavy, with tapered ends interspersed with collagen fibers. However, focal areas in high-power magnification showed atypical cells with hyperchromatic and pleomorphic nuclei without abnormal mitosis (**Figures 7-9**). Tumor cells showed strong and diffuse positivity for S-100 in immunohistochemical testing.

Microscopic examination of benign mesenchymal neoplasm excluded salivary gland neoplasms. Microscopic features along with IHC excluded other benign mesenchymal neoplasms and confirmed the diagnosis of schwannoma. The cytologic and degenerative changes supported a diagnosis of ancient schwannoma.

Discussion

To best of our knowledge, this is the first reported case of oral ancient schwannoma in Saudi Arabia. Interestingly, most of the reported cases were in Asia, accounting for two-thirds of the reported cases globally. India and Iran represent more than one-third of reported oral ancient schwannoma cases [4].

Schwannomas are typically asymptomatic, although pain and discomfort can accompany the tumor, particularly when it involves submucosal areas. The cause of schwannoma is still elusive [5]. All of the reported cases of oral ancient schwannomas were accompanied by pain with or without paresthesia are 4 cases, and all in females; 3 out of 4 of these cases were located in the mandible. Only 1 case was in the upper lip and demonstrated intermittent paresthesia [6-9], which might indicate that the intraosseous lesions are inclined to be painful. Pressure of the neoplasm on the associated nerve, which is fully surrounded by bone, may be the primary reason. In addition, females were more likely to show symptoms than males. The lesion was painless in the current case.

Other types of symptoms also have been documented. Two reported cases (both males) had difficulty swallowing or speaking; they were located in the tip of the tongue and submandibular area. The size of the tumor can be the key factor influencing symptoms: the size of the lesions in these 2 cases were 3 cm and 7.5 cm, respectively [10,11].

The floor of the mouth is the most common location of oral ancient schwannoma. Other affected sites include the tongue, jawbone, palate, submandibular area, and labial and buccal mucosa [4]. The current lesion was located in the submucosal area of the buccal mucosa, and only 2 previously reported cases had lesions at this location; both were 2 cm in diameter in 66- and 60-year-old patients. The durations of these 2 lesions were 13 and 23 years, respectively, without any complications [12,13].

There are 5 microscopic categories of schwannomas: common, plexiform, cellular, epithelioid, and ancient. The neck is the most common location of ancient schwannomas. In the oral cavity, they tend to occur in the floor of the mouth [10].

Alotaiby et al evaluated all neural neoplasms in the oral cavity, which represent 0.2% of all oral specimens. Ancient schwannomas account for 0.7% of all oral neural neoplasms, reflecting their extreme scarcity [3].

Intra-oral ancient schwannoma is an uncommon subtype of schwannoma that has some features of conventional schwannoma, including a capsule and being well demarcated from the surrounding tissues. Hyalinization, hemorrhage, cyst formation,

Indexed in: [PMC] [PubMed] [Emerging Sources Citation Index (ESCI)] [Web of Science by Clarivate]

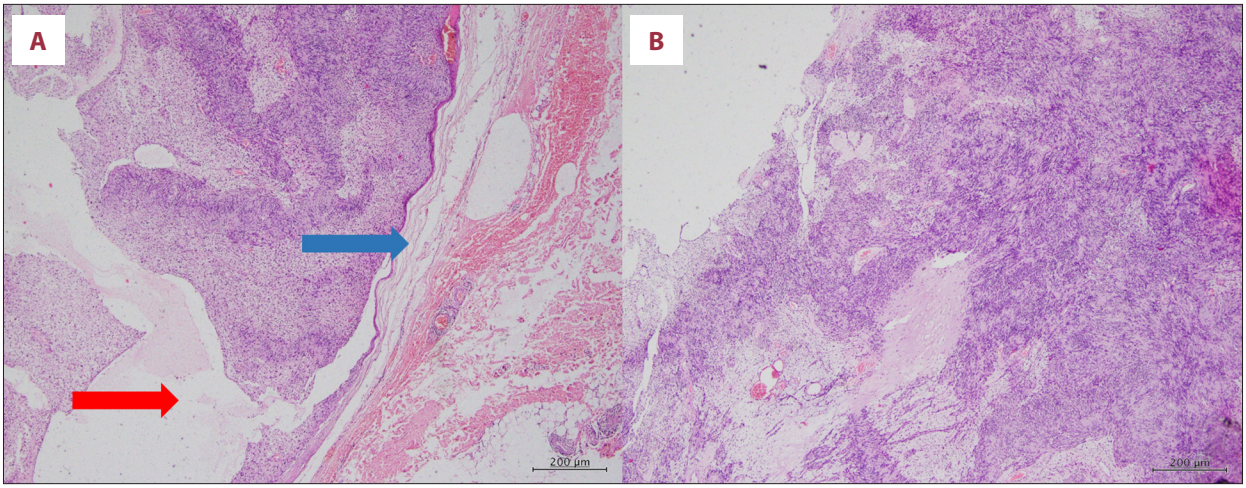

Figure 7. (**A, B**) Low-power magnification of H&E (4×) sections of ancient schwannoma showing a capsule around the lesion (blue arrow) and cystic spaces within the lesion (red arrow).

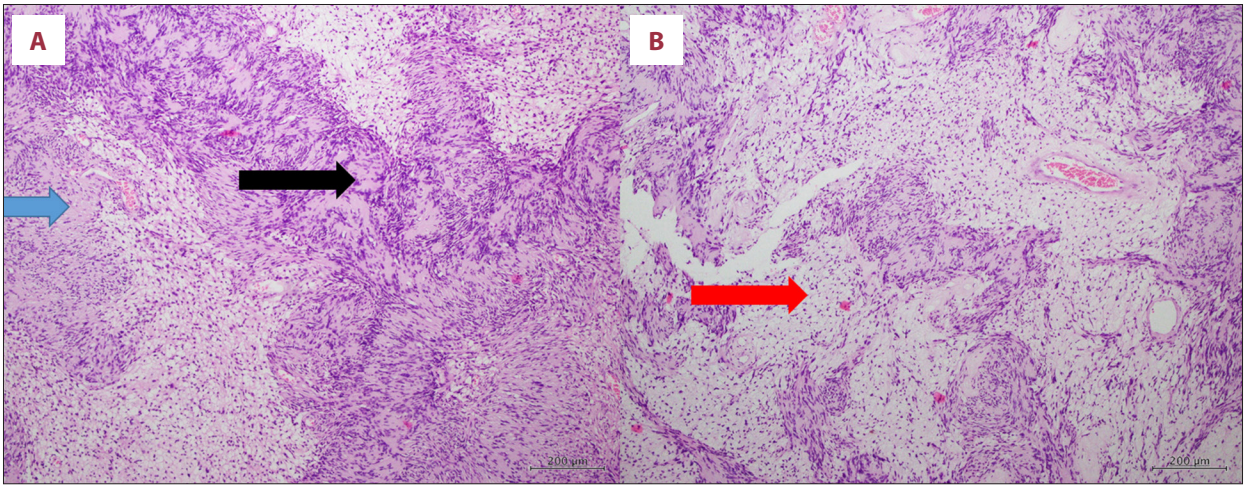

Figure 8. (**A, B**) Medium-power magnification of H&E (10×) sections show hyalinization (blue arrow) and myxomatous background (red arrow) with formation of blood vessels. Some areas show classic features of schwannoma, including Antoni A and Antoni B, as well as Verocay bodies (black arrow).

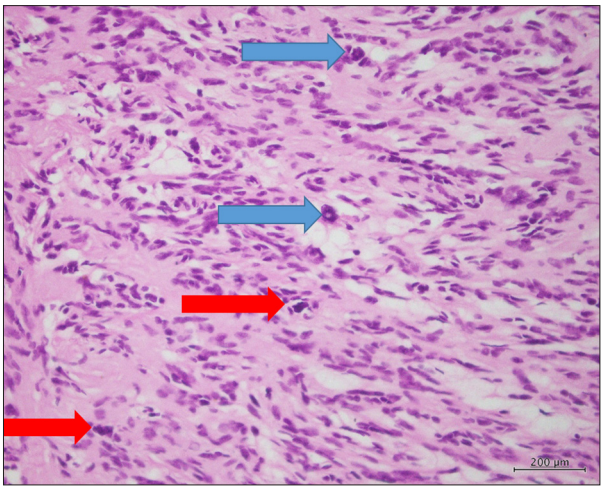

Figure 9. High-power magnification of H&E (20×) section shows pleomorphism (blue arrows) and hyperchromatism (red arrows) without mitosis.

histiocytes and siderophages infiltration and calcification are among the degenerative processes that characterize it. Some authors relate it with long history as well [10,12] In the current case, most of the microscopic features of the ancient schwannoma were noticed. However, the history of the lesion is not long, 1-2 years. The microscopic features may not necessarily reflect old lesion age.

Salehinejad et al, found that females are affected twice as often as males, with an age range of 11-82 years and an average age of 40 years. Interestingly, most of the reported cases were in Asia [4], which may suggest genetic or environmental

Indexed in: [PMC] [PubMed] [Emerging Sources Citation Index (ESCI)] [Web of Science by Clarivate]

factors associated with development of the tumor with such degenerative or ancient changes.

The treatment of schwannoma is complete surgical excision, keeping in consideration preservation of the related nerve [14-16]. The reported incidence of postoperative paralysis in vocal cord is as high as 85% in some series [17]. Harazono et al reported no recurrence in a 20-year retrospective study of surgically excised ancient schwannomas in the maxillofacial region [18]. In the present case, the surgery was done uneventfully under local anesthesia. The patient was followed up for 1 year, with no evidence of recurrence or complications.

Conclusions

We report the first case of oral ancient schwannoma in Saudi Arabia and add 1 more case to the literature. Reporting of such an unusual case familiarizes oral pathologists with histologic degenerative changes and cytologic atypia of oral ancient schwannoma. This may limit the likelihood of misdiagnosis and unnecessary treatment of this benign tumor, with an excellent prognosis.

Department and Institution Where Work Was Done

Department of Maxillofacial Surgery and Diagnostic Sciences, College of Dentistry, Qassim University, Qassim, Kingdom of Saudi Arabia.

Declaration of Figures' Authenticity

All figures submitted have been created by the authors who confirm that the images are original with no duplication and have not been previously published in whole or in part.

References:

1. Salla JT, Johann AC, Garcia BG, et al. Retrospective analysis of oral peripheral nerve sheath tumors in Brazilians. Braz Oral Res. 2009;23:43-48
2. do Nascimento GJ, de Albuquerque Pires Rocha D, Galvão HC, de Lisboa Lopes Costa A, de Souza LB. A 38-year review of oral schwannomas and neurofibromas in a Brazilian population: Clinical, histopathological and immunohistochemical study. Clin Oral Investig. 2011;15:329-35
3. Alotaiby FM, Fitzpatrick S, Upadhyaya J, et al. Demographic, clinical and histopathological features of oral neural neoplasms: A retrospective study. Head Neck Pathol. 2019;13(2):208-14
4. Salehinejad J, Sahebnasagh Z, Saghafi S, et al. Intraoral ancient schwannoma: A systematic review of the case reports. Dent Res J (Isfahan). 2017;14(2):87-96
5. Enoz M, Suoglu Y, Ilhan R. Lingual schwannoma. J Cancer Res Ther. 2006;2(2):76-78
6. Kim NR, Chung DH, Park DS, et al. Ancient schwannoma in oral cavity: A report of two cases. J Korean Assoc Oral Maxillofac Surg. 2011;37:530-34
7. Salehinejad J, Babazadeh F, Saghafi S, et al. Intra-osseous degenerated neurilemmoma of the mandible in a 23-year-old woman. J Mashhad Dent Sch. 2010;33:353-60
8. Jahanshahi G, Haghighat A, Azmoodeh F. Intraosseous neurilemmoma of the mandible: Report of a rare ancient type. Dent Res J (Isfahan). 2011;8:150-53
9. Humber CC, Copete MA, Hohn FI. Ancient schwannoma of upper lip: Case report with distinct histologic features and review of the literature. J Oral Maxillofac Surg. 2011;69:e118-22
10. Sayed SI, Rane P, Deshmukh A, et al. Ancient schwannoma of the parapharynx causing dysphagia: A rare entity. Ann R Coll Surg Engl. 2012;94(7):e217-20
11. Bilici S, Akpinar M, Yigit O, Günver F. Ancient schwannoma of the tongue: A case report. Kulak Burun Bogaz Ihtis Derg. 2011;21:234-36
12. Tobita T, Ikeda H, Fujita S, et al. Ancient schwannoma in the buccal region. Asian J Oral Maxillofac Surg. 2008;20:139-43
13. Kim NR, Chung DH, Park DS, et al. Ancient schwannoma in oral cavity: A report of two cases. J Korean Assoc Oral Maxillofac Surg. 2011;37:530-34
14. Krishnamurthy A, Ramshankar V, Majhi U. Ancient cervical vagal schwannoma: A diagnostic challenge. Indian J Surg Oncol. 2013;4(3):284-86
15. Gibber MJ, Zevallos JP, Urken ML. Enucleation of vagal nerve schwannoma using intraoperative nerve monitoring. Laryngoscope. 2012;122(4):790-92
16. Nakano CG, Massarollo LC, Volpi EM, et al. Ancient schwannoma of the vagus nerve, resection with continuous monitoring of the inferior laryngeal nerve. Braz J Otorhinolaryngol. 2008;74(2):316
17. Chiofalo MG, Longo F, Marone U, et al. Cervical vagal schwannoma. A case report. Acta Otorhinolaryngol Ital. 2009;29(1):33-35
18. Harazono Y, Kayamori K, Sakamoto J, et al. Retrospective analysis of schwannoma in the oral and maxillofacial region: Clinicopathological characteristics and specific pathology of ancient change. Br J Oral Maxillofac Surg. 2022;60(3):326-31

Indexed in: [PMC] [PubMed] [Emerging Sources Citation Index (ESCI)] [Web of Science by Clarivate]