Ancient schwannoma of the infratemporal fossa

Saravanan Balasubramaniam, Sethurajan Balasubramanian, Rohini Thirunavukkarasu
Department of Oral and Maxillofacial Surgery, Tamil Nadu Government Dental College and Hospital, Chennai, Tamil Nadu, India

Address for correspondence:
Dr. Saravanan Balasubramaniam,
35-A, Bharathi Salai, Triplicane, Chennai - 600 005,
Tamil Nadu, India.
E-mail: devasara17@yahoo.co.in

INTRODUCTION

Schwannomas are benign neurogenic tumors originating from the Schwann cells of peripheral, cranial and autonomic nerves. Though schwannomas constitute 25-40% of head and neck tumors, the incidence of intraoral schwannomas is merely 1%. Among the intraoral schwannoma the occurrence of the variant “ancient schwannoma of the infratemporal fossa with an intraoral presentation is very rare. The purpose of this presentation is to highlight the rarity of this lesion, to stress the importance of an accurate diagnosis and to consider neurogenic lesions in the differential diagnosis of head and neck lesions.

Keywords: Ancient schwannoma, infratemporal fossa, schwannoma

ABSTRACT

Schwannomas are benign tumors of neurogenic origin arising from the Schwann cells of peripheral, cranial, and autonomic nerves. Schwannomas in spite of constituting 25-40% of head and neck tumors the percentage of intraoral schwannomas is merely 1%. Among the intraoral schwannoma the occurrence of the variant “ancient schwannoma of the infratemporal fossa with an intraoral presentation is very rare. The purpose of this presentation is to highlight the rarity of this lesion, to stress the importance of an accurate diagnosis and to consider neurogenic lesions in the differential diagnosis of head and neck lesions.

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INTRODUCTION

Schwannomas are benign neurogenic tumors originating from the Schwann cells of peripheral, cranial and autonomic nerves. Though schwannomas constitute 25-40% of head and neck tumors,[1] the incidence of intraoral schwannomas is merely 1% with tongue being the most common site.[2] Schwannomas are slow growing, solitary, asymptomatic encapsulated lesions. Histopathologically there are many variants of schwannomas such as common, glandular, plexiform, cellular, epithelioid, melanotic and the ancient schwannomas.[3]

The term ancient schwannoma was coined by Ackerman and Taylor for schwannomas of long duration.[4] The first case of ancient schwannoma of the oral cavity was reported by Eversole and Howell in the floor of the mouth and on the ventral surface of the tongue.[5] In this report, a rare case of variant of schwannoma in the infratemporal fossa with an intraoral extension in the buccal mucosa has been discussed.

CASE REPORT

A 40-year-old woman was referred to the Department of Oral and Maxillofacial Surgery with the complaint of a painless swelling in the left cheek region with duration of 8 months [Figure 1a and b]. The patient revealed that she had got her left upper molars extracted 2 months prior to her presentation to us. The swelling was initially small and had gradually increased to the present size. No neurological deficit was evident on subjective and objective examination. On extraoral examination, an ill-defined diffuse swelling of 6 × 5 cm size over the left cheek region was seen extending superiorly to the infraorbital region, and inferiorly 1 cm lateral to the angle of the mouth. The swelling extended medially till the ala of the nose and laterally to the preauricular region. It was firm to palpation and was not fixed to the underlying structures. There was no associated lymph node enlargement.

On intraoral examination, a diffuse swelling of 6 × 5 cm in size was noted on the left buccal mucosa, which extended superiorly until the maxillary tuberosity with obliteration of the buccal sulcus in relation to left maxillary molars. The lesion was firm in consistency with a smooth surface, and the overlying mucosa was normal. The lesion was evaluated with a panoramic radiograph and subsequently with a computed tomography (CT) scan. The panoramic radiograph showed destruction of the alveolar bone corresponding to the extracted molar region. The CT scan showed a hypodense lesion involving the infratemporal fossa [Figure 2a-c]. It was interpreted as a benign mesenchymal tumor.

An incisional biopsy was performed and submitted for histopathological evaluation. The report revealed interlacing fascicles of spindle cells with dark staining wavy nuclei with less prominent amorphous eosinophilic zones representing verocay
bodies and perivascular hyalinization. The cellular proliferations were intermixed with large foamy histiocytes along with areas of hemorrhage and cystic degeneration [Figure 3a-c]. Although in certain fields there were nuclear hyperchromatism and mitosis, the overall histological architecture was consistent with a diagnosis of ancient schwannoma.

In keeping the indolent nature and imaging findings, surgical excision of the lesion was planned through a transantral approach to the infratemporal fossa. A Weber-Fergusson incision was made, and the maxillary bone over the lesion was exposed till the pterygoid plates. The lesion was dissected from the buccal mucosa and traced to infratemporal fossa through transantral approach and was excised in toto. The immediate postoperative period was uneventful. The surgical specimen was submitted for histopathological evaluation, which confirmed the diagnosis of ancient schwannoma [Figure 4a-d]. The patient is periodically monitored and is disease free at the last follow-up (2 years after surgery) [Figure 5a and b].

DISCUSSION

Schwannomas are benign encapsulated neurogenic lesions originating from the nerve sheath of Schwann cells occurring along the course of peripheral, cranial, or sympathetic nerve.[6] Although approximately 25-40% of all schwannomas occur in the head and neck region, the incidence of ancient schwannoma in this region is very rare. Schwannomas are designated as ancient when there is either cystic or fatty degeneration, focal accumulations of hyaline material, thick capsule, infiltration of histiocytes, siderophages and hyperchromatism. These histopathological features were attributed to the long duration of the schwannoma.[4]

Ancient schwannoma usually present as a submucosal swellings with a higher female predilection.[7] A review reveals that only nine cases of intraoral ancient schwannoma has been reported with five in the floor of the mouth and one each in the posterior maxillary mucobuccal fold, anterior maxillary vestibule, and the posterior mandibular vestibule and upper lip.[8] It is not uncommon for schwannoma to involve the infratemporal fossa, but there has been not much published reports of ancient schwannoma in this location (infratemporal fossa).[8,9] The tumors of the infratemporal fossa may arise as a primary lesion, or extend from adjacent structures or represent a metastasis. However, the literature indicates that it often occurs due to the extension from the neighboring structures.[10] In a similar context, the presenting lesion might have been a primary lesion in the infratemporal fossa with subsequent intraoral extension or vice versa. The origin of ancient schwannoma in this lesion can be traced to the trigeminal nerve, most probably the maxillary nerve due to its manifestation in the infratemporal fossa.

Unlike schwannoma, the CT findings of ancient schwannoma present as a well-defined hypodense lesion indicating the presence of cystic changes in an otherwise solid tumor similar to the presenting lesion.[11]

Schwannoma is characterized histologically by two patterns, Antoni type A where the closely packed Schwann cells are arranged in bundles and Antoni type B in which the loosely arranged Schwann cells are set within a meshwork of reticulum fibers along with verocay bodies with a well-defined capsule.[12]

According to Ackerman and Taylor, ancient schwannoma in addition typically show nuclear atypia, cystic degeneration, hyperchromatism, xanthomatous change, and fibrosis. The
current case meets these features for the diagnosis of ancient schwannoma. However, the densely cellular nature of spindle cells with hyperchromatism and mitosis together with inconspicuous Verocay bodies and abundance of foamy macrophages may lead to an erroneous diagnosis of malignancy, especially in a limited biopsy sample. This pitfall was emphasized by Dahl in his review of 11 cases of ancient schwannoma in which 4 cases have been misdiagnosed as soft tissue sarcomas due the presence of nuclear atypia and hyperchromatism. Therefore, caution should be exercised to interpret the pathology report in the context of clinical and imaging details.

Ancient schwannoma being a benign lesion has a favorable prognosis and, therefore, a conservative surgical excision would suffice with a nil recurrence rate. Though there are various surgical approaches to access lesions in the infratemporal fossa, it is most often the histopathological nature and the extent of the lesion that decides the approach, but at the same instance the decided approach should provide maximal exposure of the lesion and minimal morbidity to the patient. In the reported case since, the lesion was traced from the buccal mucosa to the infratemporal fossa the use of a Weber-Fergusson incision with transantral approach to the infratemporal aspect of the lesion is justified. The chances of malignant transformation of ancient schwannoma is rare however there are reported cases in the literature of ancient schwannoma being able to decease the pathologist of malignancy and in turn the surgeon. It is imperative to perform an adequate biopsy of any lesion, and an open mind is essential before arriving at a diagnosis for proper execution of the planned treatment.

CONCLUSION

We have presented a unique case of ancient schwannoma of infratemporal fossa to stress the rarity of the lesion and the importance of not ruling out neurogenic tumors in the diagnosis of head and neck lesions. A careful preoperative clinical and histopathological evaluation and correlation is essential as the ancient schwannoma can deceive the pathologist of a malignancy and in turn the surgeon. It is imperative to perform an adequate biopsy of any lesion, and an open mind is essential before arriving at a diagnosis for proper execution of the planned treatment.

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