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A Rare Case of Ancient Schwannoma on the Tip of the Tongue

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Abstract

Background: Neurilemmoma, neurinoma, perineural fibroblastoma or Schwann cell tumour, is a rare encapsulated benign tumour originates from schwann cells and it is of unknown etiology. About 11% - 45% of all schwannomas are found in the head and neck. The tumour can occur anywhere from the base of the skull to the thoracic inlet, but is commonly seen in the mid neck. It can rarely be seen orally as it makes up 0.04% of all intraoral tumours. *Case information*: We present a 60-year-old female complained of a 15-year-old tumour on the tip and right border of the tongue. *Methods*: Histopathological and immunohistochemistry tests for both incisional and excisional biopsies were examined for a definitive diagnosis, because clinically the tumour appears similar to reactive and other soft tissue neoplasms. *Results*: Surgical excision with safety margin done under general anesthesia without recurrence. A diathermy was used to mark and coagulate the incision. *Conclusion*: Ancient schwannoma is a rare benign encapsulated tumour of long duration. Its diagnosis is confirmed by histopathological and immunohistochemistry testing with S100 and vimentin. A magnetic resonance imaging is the choice of imaging for an accurate measurement of the tumour. Ancient schwannoma is treated surgically with total pericapsular surgical excision of the tumour and it has excellent outcomes and no recurrence.

Keywords

Tongue Tumour, Neurilemmoma, Neurinoma, Perineural Fibroblastoma, Schwann Cell Tumour, Lingual Schwannoma, Ancient Schwannoma

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1. Introduction

This benign rare encapsulated tumour originates from Schwann cells [1-27], therefore it may arise from cranial and spinal nerve roots or peripheral nerves but not from optic or olfactory nerves (lack of Schwann cells) [28]. These cells coat the nerves to augment nerve conduction and are developed embryologically in the 4th week from specialised ectomesenchymal cells of neural crest [29]. The tumour may arise from the glossopharyngeal, vagus, accessory and hypoglossal, the sympathetic chain and laterally from the cervical or branchial plexus. However, it has a predilection for the eighth cranial nerve, the vestibulocochlear nerve [5, 11, 18, 24, 27, 30]. It also has a predilection for the sensory nerves but a motor nerve may also be affected [30].

Schwannomas can rarely be seen orally (1%) [1, 2, 19, 20, 23, 26, 31-37]. They are frequently seen on the scalp, face,

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pharynx, parotid gland, middle ear, external auditory meatus, upper or lower extremities (flexor surfaces), mediastinum or peritoneum but 11% - 45% of all schwannomas are found in the head and neck [5, 8, 16] and 1%-12% of head and neck tumours may be intraoral [6, 21, 24, 27, 30, 31, 38, 39]. Rarely, a case such as a schwannoma on the lateral cervical region of the neck has been reported [38]. Intraorally, the tongue is the most common site followed by the buccal mucosa, medullary bone of the maxilla and mandible, floor of the mouth, gingiva, vestibular mucosa, lips [3, 4, 25, 29, 34, 35] and palate [1, 34, 35]. The oropharynx is a rare site but has been reported [3, 4, 29].

The tumour has no genetic and no age predilection [4, 23, 40, 41]. It has been rarely reported intraosseously [17, 21, 31]. Some articles claim no sex predilection [4, 9, 20, 24, 25, 27, 33, 37, 41, 42], but Gainza-Cirauqui et al. stated the female to male ratio as 1.6:1 [4, 19, 21, 31]. It appears clinically similar to reactive and other soft tissue neoplasms [2, 9, 10, 15, 32-35, 43]; a slowly growing, painless, well circumscribed [6, 9, 10, 19, 22, 25, 34-36, 41, 42], exophytic, and nonindurated swelling [4, 32]. Here we report a case of an Ancient Schwannoma on the tip of the tongue.

2. Case Report

A 60-year-old female complained of a 15-year-old lump on the right tip of her tongue with an ulcer. It affected her speech and was irritating when she chewed. She did not mention any history of symptoms of von Recklinghausen's disease or of a chronic illness. She sought medical attention a week before she came to our clinic, where she was referred under the impression that it might be tongue carcinoma because of the ulcer.

The capsulated lump (3x2cm) was smooth except for the side adjacent to the canines and premolars (upper and lower) which was ulcerated. The tongue movement was not restricted with no change in sensation or taste. No lymph nodes were palpable or tender.

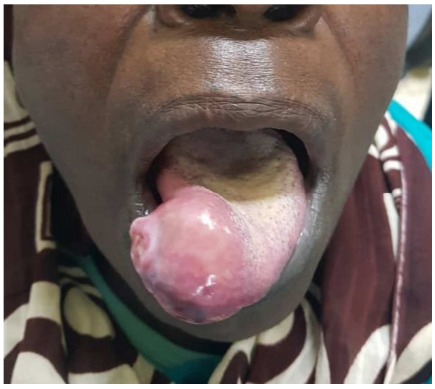

Figure 1. Anterior view of the lesion.

Figure 2. Superior view of the lesion.

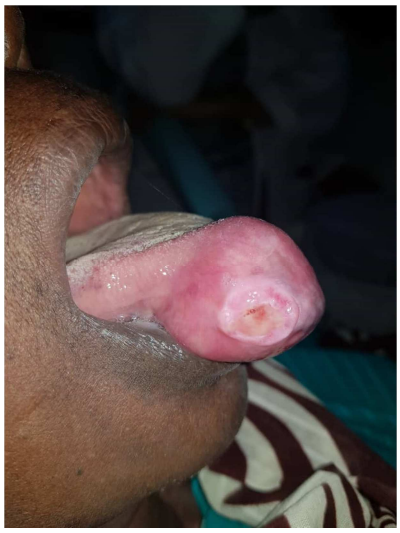

Figure 3. Lateral view of the lesion.

Figure 4. Anterio-lateral view of the lesion.

An incisional biopsy was done including the ulcerated site. Intraoperatively a viscous straw-coloured fluid was discharged as soon as an incision was made into the capsule. Section showed a benign lesion composed of sheets of clear cells with peripheral nuclei and spindle cells with collagen production, highly suggestive of spindle cell lipoma. No immunohistochemistry was done to confirm the diagnosis.

Excisional biopsy under GA was decided, given the rich blood supply of the tongue. A diathermy was used to mark

and coagulate the incision with a safety margin of 0.5cm. The bleeding was controlled and the tongue sutured and recontoured after releasing the mucosa.

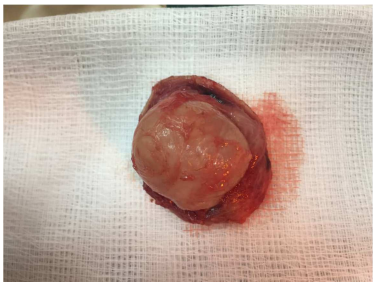

Figure 5. The Excisional biopsy.

The histopathological report revealed small spindle cells with serpentine nuclei and a focal area showing Verocay bodies resembling Antoni A regions, large areas with degenerative changes and numerous cystic spaces with haemorrhage surrounded by numerous nerve bundles. An immunohistochemistry test for S100 was positive, highly suggestive of ancient schwannoma.

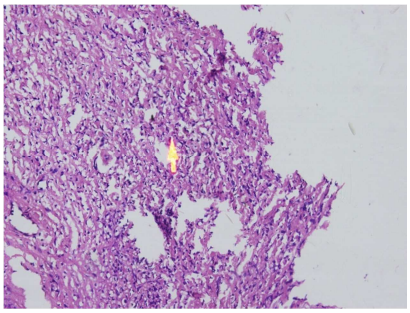

Figure 6. Spindle shaped Schwann cells.

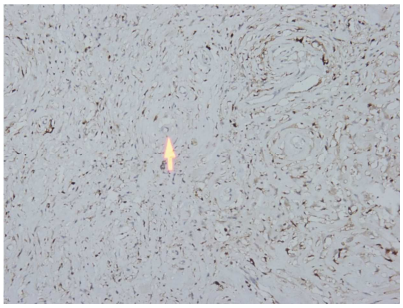

Figure 7. S100 positive Schwann cells.

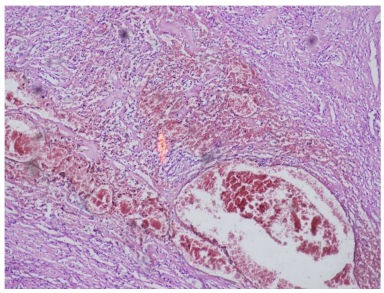

Figure 8. Haemorrhagic changes.

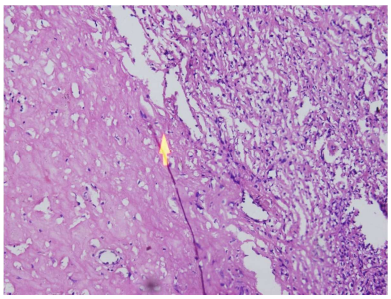

Figure 9. Degenerative changes.

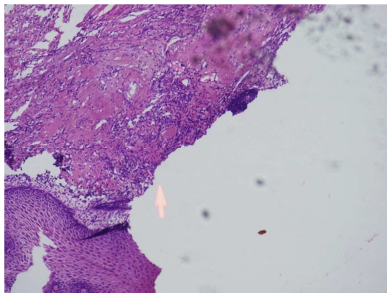

Figure 10. Verocay bodies and Schwann cells.

3. Discussion

Schwannoma was first described by Verocay in 1910 [44] (some case reports stated 1907 [45]) and he named it neurinoma [39]. In 1932, Masson introduced ‘schwannoma’ [31] and in 1935 Stout called it neurilemmoma [10]. It is also called perineural fibroblastoma [23, 39]. Eversole and Howell (1971) were the first to report an intraoral case of ancient schwannoma in detail [46] but the term ‘ancient’ was introduced by Ackerman and Taylor [3, 5, 7, 12-15, 17, 35, 47, 48].

The rare tumour makes up 0.04% of all intraoral tumours [49] and 1%-12% of head and neck tumours [30]. Cranial nerves are affected, mostly the vestibulocochlear nerve. Cases arising from hypoglossal [50], lingual [3] and mental [51] nerves have been reported.

Fifty percent of the cases can be linked directly to a nerve [29, 30]. The vagus nerve is said to be the origin of 50% of parapharyngeal schwannomas followed by the cervical sympathetic chain. In this case, the origin is most likely a peripheral root of the hypoglossal [4] or lingual nerve. The tongue is the most common site intraorally [1, 6, 23, 24], which represent 1% of all head and neck schwannomas [38]. Two thirds of these lingual cases occur in the anterior two thirds [2, 43]. Tongue lesions located at one side of the tongue represent 36.3%, followed by 24.2% at the tongue base, 21.2% in the tip and 15.1% in the ventral surface, according to Erkul et al. [38]. Although intraorally schwannomas are more common on the tongue; ancient schwannomas are found less intraorally and located more on the floor of the mouth [4]. Intraosseous, although rare, manifests with pain and paresthesia with bone expansion

usually in the posterior mandible [4, 17]. Given the site of the tumour in this case, she did not show any symptoms such as pain, dysphagia, snoring, otalgia or change of voice [40]. She complained of the awkward speech and difficulty in chewing food.

As there is no age predilection, some articles claim that schwannomas are more common in between third and fifth decayed of life [1, 5, 7, 8, 10, 13, 27, 33], and older for ancient schwannoma [4]. This case study's age is at the end of the spectrum for ancient schwannomas, however the gender is consistent with the predilection [1, 5, 8, 18, 31, 34, 36], although most literature states no predilection to gender or race [13, 34, 52].

The etiology of this tumour is unknown [4, 23-25, 36, 39]. Hwang et. al [28] and Lee et. al reported the case to be a tongue bite [38]. Our patient said she couldn't recall any sort of trauma to her tongue. Schwann cells tumours are found accidentally after being inactive for a long period [30, 40]. As in this case, it took her 15 years before she sought medical treatment.

In relation to the size, oral ancient schwannomas are larger than conventional schwannomas and can range from less than 1cm up to 7cm [4]. Orally, there are two types:

(1) Submucosal – encapsulated, well-defined, firm (resembling a cyst)

(2) Non-capsulated – found below the basal layer of the mucous membrane [3, 4, 53].

Histopathological and immunohistochemistry tests are essential for a definitive diagnosis [11], because clinically the tumour resembles lipoma, fibroma, leiomyoma or adenoma [42, 54]. Its characteristic features include:

(1) Encapsulated mass with degenerative alterations containing both large cystic and solid areas

(2) A mixture of spindle cells with highly cellular (Antoni A) and less cellular myxoid (Antoni B)

(3) Palisaded nuclear appearance of Schwann cells

(4) Verocay bodies seen in a cellular eosinophilic zone [3, 4, 6, 7, 12, 17, 21, 22, 26, 27, 34, 36, 39].

All of the above were seen in the histological specimen. Furthermore, the immunohistochemistry test for S100 was positive, further confirming the diagnosis [5, 7, 8, 11, 17-19, 21, 22, 25-27, 34, 35, 39, 41, 50]. The specimen may also be tested for vimentin, Leu-7 antigen and glial fibrillary acidic protein, and in the case it is an ancient schwannoma the test should be positive [2]. The name 'ancient' conveys the long duration of the tumour which is seen in the degenerative changes [5, 7, 8, 13, 14, 17, 39]. A theory states the size of the tumour creates vascular insufficiency and thus the degenerative changes [7, 13, 47].

When in doubt or when the lesion is difficult to assess, such as those on the base of the tongue or oropharynx, an MRI is the choice of imaging [4, 7, 8, 14, 15, 19, 22, 26, 27, 37, 40]. It is superior to a CT scan [27, 30] in that it is not degraded by dental amalgam or beam- hardening artefacts and provides a precise measurement of the tumour. On MRI, tongue schwannomas appear isointense to muscle on T1-weighted images and homogenously hyperintense on T2-weighted images [8, 9, 13, 26-28, 31, 38, 39, 42, 45, 47, 54].

Total pericapsular surgical excision of the tumour is the first line of treatment and it has excellent outcomes [1, 2, 4, 7, 8, 10, 13, 14, 17, 19, 22, 23, 25, 26, 29-39, 43, 50] and no recurrence [29, 38]. Nevertheless, complications are present and may vary from scarring, anesthetic complication, loss of tongue function, airway management [40] and Horner's syndrome [8, 12]. A diagnosis of malignancy transformation has been mentioned in 8%-10% of the reported cases [2, 30]. In the literature which has been reviewed, one case was reported for recurrence after surgical excision 23 years ago. The recurring tumour was bilobular, one of which was believed to be a malignant nerve sheath tumour [55]. In this case presented, there were no signs of recurrence for up to 3 years postoperatively. For the diagnosis of malignant transformation to be made, the tumour should demonstrate in part a benign schwannoma, unequivocal malignant foci, transitional area between the benign and malignant schwannoma and the patient should have no evidence of von Recklinghausen's disease [29].

4. Conclusion

Ancient schwannoma is a rare benign encapsulated tumour of long duration, either found accidentally or the patient notices it as it expands. Age has no predilection. The female gender is slightly more prone to the tumour than the male. It should be considered among the differential diagnoses and confirmed by histopathological and immunohistochemistry testing with S100 and vimentin. MRI is the imaging modality of choice for an accurate measurement of the tumour. Surgical excision of the lesion with preserving adjacent vital structures is effective with a good prognosis. Recurrence and malignant transformations are rare.

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