



A rare case of ancient schwannoma of the tongue



Ann Ital Chir, Digital Edition 2020, 9
pii: S2239253X20033897 - Epub, September 14
free reading: www.annitalchir.com

Ida Barca*, Daniela Novembre*, Kallaverja Elvis*, Valeria Zuccalà**, Maria Giulia Cristofaro*

*Department of Experimental and Clinical Medicine, Unit of Maxillofacial Surgery (Chairman: Prof.ssa M.G. Cristofaro) "Magna Graecia" University, Catanzaro, Italy

**Pathology Unit, Pugliese-Ciaccio Hospital, Catanzaro, Italy

A rare case of ancient schwannoma of the tongue

AIM: This is a case of Ancient schwannoma, an uncommon schwannoma variant, of clinical interest for the unusual site of the lesion, the diagnostic complexity and the surgical approach.

CASE REPORT : We present a 40-year-old patient with intramural swelling of the lingual belly. Surgical removal was performed, and the diagnosis was confirmed by histopathological examination.

DISCUSSION: We discuss the clinical findings and therapeutic strategies for treating and diagnosing ancient schwannoma.

CONCLUSIONS: The specific diagnosis is difficult and frequently late. Growth is slow and only at a distance of time symptoms arise. These tumors must be treated surgically in the prevention of a possible recurrence.

KEY WORDS: Ancient schwannoma, Intramural tongue mass, Antoni A and Antoni B bodies, Neurinoma, Neurilemoma

Introduction

Schwannoma, also called Neurinoma or Neurilemoma, is a benign neof ormation that derives from the Schwann cells (glial cell complex) that form the sheath of the peripheral nerves and is one of the most frequent tumors of these nerves and rarely occurs for in the head and neck region. They appear as slow-growing, ovoid and well-circumscribed masses with a minimal probability of degenerating. Clinically it is asymptomatic, without pain and without compromising the nerves. The etiology of these lesions remains unknown. The etiology of these lesions remains unknown. Sporadic cases have been

attributed to previous traumatic events or chronic irradiation ¹. Ancient Schwannoma is a variant which microscopically can show an atypical nuclear degeneration that should be confused with a malignant lesion ². The treatment is based on the surgical excision of the neof ormation in the prevention of a possible recurrence. We report a rare case of intra-mural Ancient Schwannoma of oral tongue in a 40-year-old patient.

Case Report

A 40-year-old male patient presents at our Maxillo-Facial Surgery Unit of the Magna Graecia University of Catanzaro, in January 2018 with limited swelling in the median intramural center of the tongue. The patient had previously been seen by the ENT team 6 months ago for otitis media and the specialist doctor had noticed the swelling in the same region. The patient did not complain of any symptoms. There was no pain, no changes in functionality and lingual movements, no deficit in sensitivity and no changes in taste. Clinically,

ervenuto in Redazione Giugno 2020. Accettato per la pubblicazione Luglio 2020

Correspondence to: Ida Barca, MD, Maxillo-Facial Surgery Unit, Department of Experimental and Clinical Medicine, "Magna Graecia", University; Viale Europa, 88100 Catanzaro, Italy; (e-mail: barca.ida@gmail.com)



Fig. 1: A well-circumscribed solitary oval swelling of about 2 x 1.5 cm in the median intramural center of the tongue at clinical presentation.

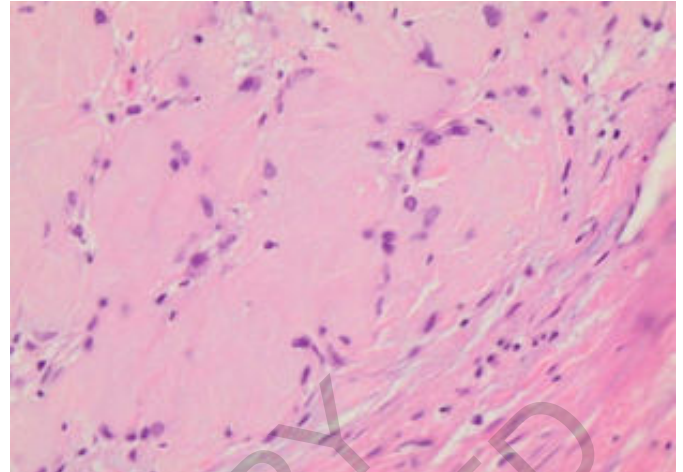


Fig. 4.: High power of schwannoma with capsule and large hyperchromatic nuclei are seen.

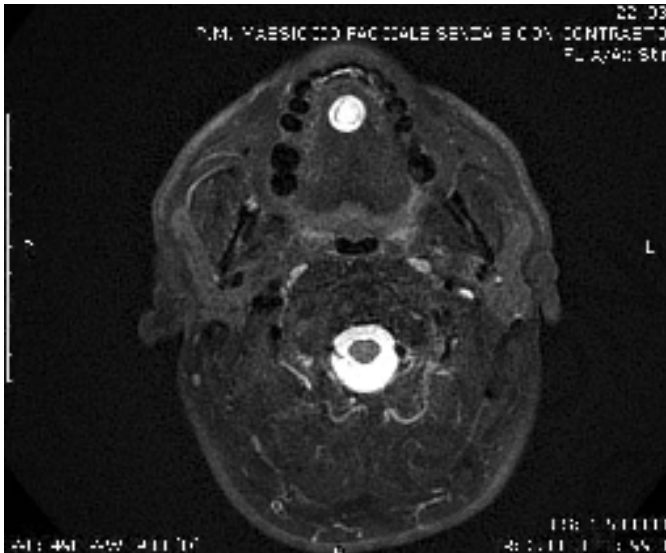


Fig. 2: Pre-operative axial STIR MRI reveals a well-defined hyperintense mass.

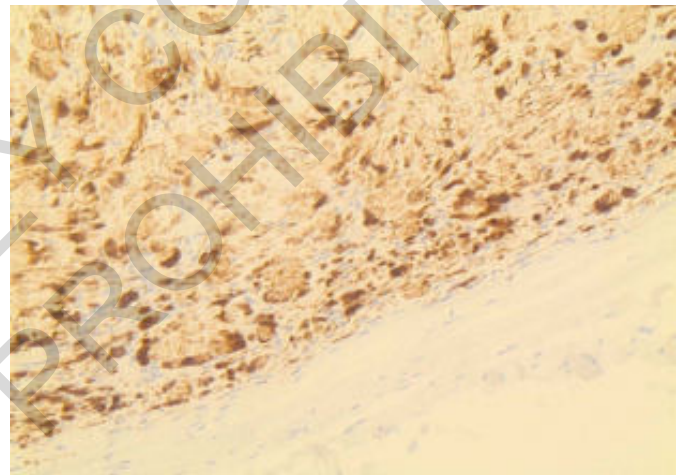


Fig. 5: S-100 protein immunoreactivity high power; large cell are seen.

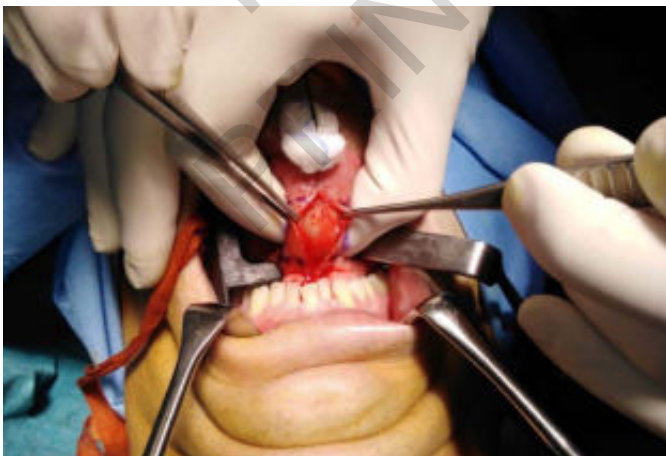


Fig. 3: The intraoperative image shows an oval capsule formation of 18 x 11 x 12 mm.



Fig. 6: Post-operative at 24 months of axial STIR MRI showed no signs of diffusivity other than signal alterations.

the lesion presented itself as a well-circumscribed solitary swelling of about 2 x 1.5 cm, oval shape, with a stretchy consistency, about 1.5 cm from the tip with extension towards the belly of the tongue and in a minimal part at the level of the back (Fig. 1). Subsequently, MRI was performed for the patient where an oval formation 18 x 11 x 12 mm isointense in T1 and in homogeneously hyperintense in T2 with hyperintense central area and after MDC central saving peripheral impregnation was found in the context of the genioglossus muscle, suspecting diagnosis of granuloma. We practiced an incisional biopsy before planning the surgery. The result was suspicious Schwannoma. During the surgery, an excision of the lesion was scheduled. Longitudinal incision was made following the course of the lingual frenulum of about 4 cm with excision of the capsule formation which was sent for histopathological examination (Fig. 3). The report showed macroscopically a greyish white lesion of 18 x 14 x 10 mm while the microscopic one showed nodular proliferation with thin capsular coating, consisting of cellular elements fused with focal nuclear pleomorphism in the absence of mitotic activity and necrosis. Cystic areas contains Antoni A and Antoni B zones. Antoni A zones are quite cellular composed of spindle cells often arranged in a palisading fashion, Antoni B zones, the tumor cells are separated by abundant edematous fluid that may form cystic space. Occasionally, isolated cells with bizarre hyperchromatic nuclei are observed (so called ancient schwannoma) (Fig. 4). Immunohistochemistry confirmed the diagnosis of consistent Schwannoma Ancient for S100, CD34, CD68, Desmina, ki 67 (Fig. 5). The patient was followed up for 6-12 and 24 months. Clinically no functional deficits in movement and / or sensitivity. Unaltered sense of taste. Control MRI was recommended at 12 and 24 months which showed no signs of diffusivity other than signal alterations (Fig. 6).

Discussion

Schwannoma (for which the terms neurinoma or neurilemmoma are also used) obviously originates from the nerve lining but involves only one or two nerve bundles; the remaining fascicles are dislocated and not invaded by the tumor, which is provided with a distinct capsule. Schwannoma has a low incidence (1/3000 births). It affects both sexes, with prevalence, according to some, of the female one, and all ages, with a slight preference for the III-IV and V decade³. Any region crossed by peripheral nerves can be a home to a schwannoma, but the most common localizations are those that correspond to the course of the nerves of the neck and skull and the surface ventral upper limbs⁴. In a percentage of cases ranging from 25 to 45% this neoplasm is localized in the cervico-cranial district. However, the most affected is the acoustic, whereas the most common site is the

parapharyngeal space originating from the cranial nerves and the cervical sympathetic chain⁵. To a lesser extent is the sympathetic cervical, glossopharyngeal, accessory, hypoglossal, and brachial plexus. In rarer cases they can involve districts such as the facial mass, external nose, paranasal sinuses, nasopharynx, oral cavity, tongue, oropharynx, vocal cords and larynx, mandibular. These lesions are almost always benign tumors, only 2% of cases have been described as having histological aspects of malignancy (cellular atypia and high number of mitosis) (2). This was first described by Verocay in 1910⁶. In the old classification that originated from the nerve sheath we distinguished:

- a) neurolemmomas: the neoformations that originate from Schwann cells of the inner layer of the lining of a nerve (neurolemma);
- b) neurilemmomas: the neoformations that originate both from the Schwann cells of the neurolemma and from the thin external membrane that surrounds the neurolemma (neurilemma).

Today these new forms are generally referred to as schwannomas⁷. Currently, peripheral nerve tumors include neurilemmomas, neurofibromas and perineural cell tumors. The WHO classification divides these tumors into benign and malignant ones where the former are: traumatic neuroma, paisadet encapsulated neuroma, hamartomas, neurofibroma / neurofibromatosis, schwannoma, granular cell tumor, nerve sheath myxoma, pigmented neuroectodermal tumor of infancy, and the malignant: malignant peripheral nerve sheath tumor, olfactory neuroblastoma, malignant granular cell tumor, peripheral neuroectodermal tumor^{8,9}. A literature review was made from 1971 until 2011 for the ancient schwannoma of the oral cavity to demonstrate the rarity of this type of tumor³.

In our study PubMed is practiced as a search engine until 2020 and an even more specific review is made in an attempt to demonstrate the uniquely lingual localization of the ancient schwannoma (Table I). During development, Schwannoma is in close relationship with the nerve of origin with the characteristic that does not creep between the nerve bundles with a well-distinct capsule that greatly helps the cleavage of the neoformation and its complete enucleation.

Macroscopically the schwannomas present themselves as unique nodules, of variable dimensions, usually less than 5 cm, of regular appearance, variable color from pink to yellow and soft consistency however, variable from soft to hard-elastic. When cut, they can present cystic areas of degeneration¹⁰. Microscopically they consist of two different components, Antoni A and Antoni B. Antoni A is an area intensely rich in spindle-shaped, ordered cells, grouped in bundles, with eosinophilic and poorly differentiated cytoplasm and basophilic nucleus. It has its characteristic appearance of the so-called "Verocay bodies", which are formed by these palisade organized cells that alternate with eosinophilic cell areas.

TABLE I - The various case reports of the literature from 1971 to 2020 are listed.

Authors (reference)	Age/Sex	Location of Schwannoma	Size	Treatment
Eversole & Howell et al. 1971 (24)	58/F	Mouth floor and ventral tongue	2.5 cm	Surgical exision
Nakayama et al. 1996 (25)	40/F	Mouth floor and ventral tongue	5.5 cm	Surgical exision
Ledesma et al. 1999 (26)	21/F	Mouth floor and ventral tongue	3.0 cm	Surgical exision
Bilici et al. 2011 (27)	45/M	Tip of the tongue	3.0 cm	Excised (local anesthesia)
Lee et al. 2013 (28)	29/ F	Lower left side of the ventral tongue	3.3 cm	Surgical exision
Shashikumar et al. 2019 (29)	35/F	Right dorsal of the tongue	5.0 cm	Surgical exision
Yaslikaya et al. 2020 (30)	32/F	Tongue base	4.5 cm	Endoscopic Transoral Resection
Current case (2020)	40/M	Ventral tongue	2.0 cm	Surgical exision

The second component (Antoni B) consists of less intensely cellular areas, rich in mixoid tissue and large thin-walled vessel, which may contain areas of degeneration. Immunohistochemical investigations are positive for the expression of the S-100 protein and for vimentin C¹¹. Both benign and malignant forms (exceptional) can have foci of epithelioid cells (squamous cells, cartilage, bone tissue, muscle tissue) in relation with the origin of the tumor from the elements of the neural crest and as a consequence of the mechanisms of interaction - induction with the mesenchyme. The diagnosis is generally late due to slow growth and silent symptomatology except in prominent cases. If less than 2 cm they are usually asymptomatic. If they exceed 3 cm, they can give symptoms such as pain, paresthesia, dislocation and compression of the original nerve. First level instrumental examinations such as ultrasound can give us an idea in the diagnosis but are often not diriment. In this case CT with contrast medium or even better an MRI is required. The typical magnetic resonance (MR) appearance is iso-to-hyperintense (compared to muscle) on T1-weighted images, hyperintense on fluid-sensitive sequences, and often diffusely enhancing on contrast-enhanced images. Tissue heterogeneity is relatively common, particularly cystic degeneration¹². When present, heterogeneity has been shown to correlate histologically with a greater ratio of Antoni B tissue than Antoni A¹³. On MR imaging (MRI), Type 1 predominant tumors tend to be small and homogeneous while heterogeneous tumors (with or without cystic degeneration) tend to have higher proportions of Type 2. Larger and more heterogeneous tumors also demonstrate increased hemosiderin deposits and may be referred to ancient Schwannomas¹⁴. A correct and early diagnosis of schwannoma presupposes the integration and complementarity of the anamnestic, clinical, instrumental examinations, intraoperative evaluation and an accurate anatomopathological, microscopic and immunohistochemical examination. The treatment always remains the surgical one which consists in the complete excision of the new formation. The interest of the case to be treated for the surgeon is technical. Surgery is planned on the basis of instrumental investigations such as MRI or CT. Surgical

resection of Schwannomas located at the base of the tongue are inherently difficult due to limited operative exposure amidst intricate neurovascular anatomy, which may cause significant morbidity when damaged, such as impaired speech, aspiration, dysarthria, and dysphagia¹⁵. Despite the fact that complications are rare this does not detract from the fact that nerve lesions in the affected region can occur.

Important to differentiate the nervous stupor that can be due to edema and / or stretching from the actual lesions that do not allow the recovery of the nerve. The results of the surgery depend on the specific experience of the surgeon, but also on a series of factors independent of the type of intervention, such as the characteristics of the lesion (size, location, previous operations, aspects of malignancy, relationships with the nerves and vessels) and operative timing. Due to the complexity of the variables involved, it is not possible to establish with certainty the time necessary for the recovery of the functions of a nerve that has manifested a deficit; the waiting period for an evaluation of the results cannot therefore be precisely defined. Radiotherapeutic treatment is not indicated as these types of tumors are resistant to ionizing radiation rather, the literature describes how radiotherapy and gamma knife (GKF) treatment could cause transformation into a malignant variant^{2,16}. Other types of experimental treatments consist in: chemotherapy (bevacizumab, mTOR kinase inhibitor, dasatinib) and CO₂ laser surgery but still insufficient data to replace the primary technique¹⁷⁻¹⁹. The prognosis is excellent in most cases. A bi – annual clinical follow up with MRI – scan control is indicated.

Conclusion

Ancient intra-murally Schwannoma represents a rare site of entity in tongue.

The early diagnosis is difficult and presupposes the integration of the anamnestic, clinical, instrumental and immunohistochemical examination. The treatment of choice is surgical excision of the tumour with excellent postoperative prognosis and rare instances of recurrence.

Riassunto

L'Ancient schwannoma è una rara variante di Schwannoma, un tumore benigno che deriva dalle cellule di Schwann (complesso di cellule gliali) che formano la guaina dei nervi periferici ed è uno dei tumori più frequenti di questi nervi che si verifica raramente nella regione testa-collo. Si presenta come una massa asintomatica, a crescita lenta, ovoidale e ben circoscritta con una probabilità minima di degenerazione. L'eziologia di queste lesioni rimane sconosciuta. Qui presentiamo il caso di un paziente di 40 anni con tumefazione intramurale del dorso linguale che è stato sottoposto ad intervento chirurgico di rimozione della lesione e la cui diagnosi è stata confermata dall'esame istopatologico. Per questi tipi di lesione a volte la diagnosi precoce risulta difficile e presuppone l'integrazione dell'esame anamnestico, clinico, strumentale e immunoistochimico.

Il trattamento di scelta è l'asportazione chirurgica del tumore con eccellente prognosi postoperatoria e rari casi di recidiva.

References

1. Sznajder L, Abrahams C, Parry DM, Gierlowski TC, Shore-Freedman E, Schneider AB: *Multiple schwannomas and meningiomas associated with irradiation in childhood*. Arch Intern Med, 1996; 156:1873-878.
2. Markou K, Eimer S, Perret C, Huchet A, Goudakos J, Liguoro D, et al: *Unique case of malignant transformation of a vestibular schwannoma after fractionated radiotherapy*. Am J Otolaryngol, 2012; 33:168-73.
3. Salehinejad J, Sahebnaasagh Z, Saghafi S, Sahebnaasagh Z, Amiri N: *Intraoral ancient schwannoma: A systematic review of the case reports*. Dent Res J (Isfahan), 2017; 14(2):87-96.
4. Kim DH, Murovich JA, Tiel RL, Moes G, Kline DG: *A series of 397 peripheral neural sheath tumors: 30-year experience at Louisiana State University Health Sciences Center*. J Neurosurg, 2005; 102:246-55.
5. Takimoto T, Katoh H, Umeda R: *Parapharyngeal schwannoma of the cervical sympathetic chain in a child*. Int J Pediatr Otorhinolaryngol, 1989; 18: 53-8.
6. Verocay J: *Zur Kenntnis der "Neurofibrome"*. Beitr Pathos Anat, 1910; 48: 1-69.
7. Enzinger FM, Weiss SW: *Benign tumors of peripheral nerves InSoft tissue tumors*. St. 3rd ed. Louis, TORONTO, London:Mosby, 1985: 821-88.
8. Allen CM, Gnepp DR, Ro CJ: *Granular cell tumor*. In: El-Naggar AK, Chan JKC, Grandis JR, Takata T, Sootweg P, eds *World Health Organization classification of head and neck tumours*. 4th edn. Lyon: IARC, 2017; Ch4; 121-22.
9. Barca I, Cordaro R, Giudice A, Cristofaro MG: *Abrikossoff's tumor of the tongue: Report of three cases and review of the literature*. J Oral Maxillofac Pathol, 2020; 24(Suppl 1): S101-S105.
10. Vered M, Fridman E, Carpenter WM, Buchner A: *Classic neurothekeoma (nerve sheath myxoma) and cellular neurothekeoma of the oral mucosa: immunohistochemical profiles*. J Oral Pathol Med, 2011; 40:174-80.
11. Santos PP, Freitas VS, Pinto LP, Freitas Rde A, de Souza LB: *Clinicopathologic analysis of 7 cases of oral schwannoma and review of the literature*. Ann Diagn Pathol, 2010; 14:235-39.
12. Murphey MD, Smith WS, Smith SE, Kransdorf MJ, Temple HT: *From the archives of the AFIP. Imaging of musculoskeletal neurogenic tumors: Radiologic-pathologic correlation*. Radiographics, 1999; 19(5):1253-80.
13. Wippold FJ 2nd, Lubner M, Perrin RJ, Lämmle M, Perry A: *Neuropathology for the neuroradiologist: Antoni A and Antoni B tissue patterns*. AJNR Am J Neuroradiol, 2007; 28(9):16338.
14. Gomez-Brouchet A, Delisle MB, Cognard C, Bonafe A, Charlet JP, Deguine O, et al.: *Vestibular schwannomas: Correlations between magnetic resonance imaging and histopathologic appearance*. Otol Neurotol, 2001; 22(1):79-86.
15. Diplan J, Cavallo P, de los Santos S: *Anterior midline glossotomy approach for large schwannoma of the tongue: Case report*. Ear Nose Throat. 2018; 11:1-3.
16. Yanamadala V, Williamson RW, Fusco DJ, Eschbacher J, Weisskopf P, Porter RW: *Malignant transformation of a vestibular schwannoma after gamma knife radiosurgery*. World Neurosurg. 2013; 79:593e1-593.e8.
17. Mautner VF, Nguyen R, Kutta H, Fuensterer C, Bokemeyer C, Hagel C et al.: *Bevacizumab induces regression of vestibular schwannomas in patients with neurofibromatosis type 2*. NeuroOncol, 2010; 12:14-18.
18. Sagers JE, Beauchamp RL, Zhang Y, Vasilijic S, Wu L, DeSouza P, et al.: *Combination therapy with mTOR kinase inhibitor and dasatinib as a novel therapeutic strategy for vestibular schwannoma*. Sci Rep, 2020; 10: 4211.
19. Mehrzad H, Persaud R, Papadimitriou N, Kaniyur S, Mochloulis G: *Schwannoma of tongue base treated with transoral carbon dioxide laser*. Lasers Med Sci., 2006; 21(04):235-37.