Original Research Paper

Volume-7 | Issue-10 | October-2017 | ISSN - 2249-555X | IF : 4.894 | IC Value : 79.96

Anatomy

NEURILEMMOMA OF SUBLINGUAL SALIVARY GLAND: A RARE CASE.

*Dr. Navneet Agarwal	Professor, Department of ENT, Dr. S.N. Medical College, Jodhpur(Rajasthan),India, *Corresponding Author
Dr. Raghav sharma	Resident Doctor, Department of Otorhinolaringology, Dr. S.N. Medical College, Jodhpur (Rajasthan), India
Dr.Ritu Agarwal	Assistant Professor, Department of and Anatomy, Dr. S.N. Medical College, Jodhpur (Rajasthan), India

ABSTRACT Neurilemmomas are well-encapsulated, benign, slow-growing tumors originating from Schwann cells of the nerve sheath surrounding cranial, peripheral, or autonomic nerves. Intraoral neurilemmomas are relatively rare (25 to 45 % of neurilemmoma occur in head and neck region out of these 20% are intra oral with tongue being most commom site). Neurilemmoma of sub lingual gland are even more rare. They have a wide variety of morphologic and radiologic features. This makes differential diagnosis difficult, and only histopathological features can lead to a definitive diagnosis. In this report, we present the case of a 55 -year-old woman who presented to Department of ENT Dr. S.N. Medical College, Jodhpur, Rajsthan, India whose chief complaint was a solitary, nodular mass in the floor of the mouth on left side. The mass was removed completely through an intraoral surgical approach. Wound healing was uneventful and there were no signs or symptoms of recurrence.

KEYWORDS : neurilemmoma, intra oral, nerve sheath, sublingual.

Introduction:-

Neurilemmomas are solitary neurogenic tumor that arise from cells of neural sheath[1]. They are slow growing and represents proliferation of schwann cells.Majority of extracranial tumors are benign . Intraoral neurilemmomas are relatively rare and have a wide variety of morphologic and radiologic features, this makes diagnosis difficult, and only histopathological features can lead to a definitive diagnosis[2]. Nearly 45% of all schwannomas occur in the head and neck area, where they may originate from any of the peripheral, cranial or autonomic nerves[1]. These benign tumors occurs in any age group with equal predilection in both sex.They are painless, insidious, slow growing tumors [3]. So, they are of long duration at the time of the presentation and rarely show a rapid course . The nerve of origin is not identified in around 10 to 40 percent of schwannomas[4]. If nerve of origin is small then the nerve of origin is not identified and if nerve of origin is large the nerve fibre are splayed in outer capsule of tumor[4]. Schwanomma of salivary gland is a rare form of extracranial neurogenic tumor. Intraoral schwannoma is a rare entity with tongue being most common site.

Schwanomma in sublingual salivary gland is extremely rare[5].

Case report:-

A 55 yr old female presented with swelling in floor of mouth for last 8 months. Initially swelling was 1 to 2 cm in size and very slowly increased in size to reach a size of 3 cm after 8 months. The patient's medical and dental history was unremarkable. No symptoms suggesting paresthesia was reported by patient. A physical examination of her floor of mouth revealed a smooth-surfaced, mobile, firm, and painless mass of 3x 2 cm in diameter.

The mass was slightly movable.The neck examination was normal and no mass or swelling was observed in the neck. No regional lymph node were palpable.

An ultrasound examination of the neck revealed a well-circumscribed and hypoechoic lesion 3.53cm x 3.15cm in size in floor of mouthmore on left side with increased vascularity.

CT Scan shows 3.8cmx3.5cmx2.5 cm well defined enhancing mass in floor of mouth more on left side abutting lingular aspect of mandible with no bony erosion.(figure 1)

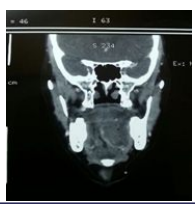

figure 1

The differential diagnosis included sublingual gland tumor and neurogenic tumors. The dilemma faced by us was weather to go for pre operative biopsy or not, since large number of sublingual tumors are found malignant. But looking at clinical and radiological features we decided to go for excision without taking pre operative biopsy.

Our patient[figure 2] underwent surgical excision of the mass under general anesthesia via intra oral route. The mass was found adherent to sublingual salivary gland with nerve of origin uncertain. So mass along with sublingual salivary gland was removed by blunt dissection and sent for histopathological examination.

figure 2

Macroscopically, the resected mass was encapsulated, yellowish in color, measuring 3cm in its greatest diameter. It was oval, smooth and firm. [figure3]

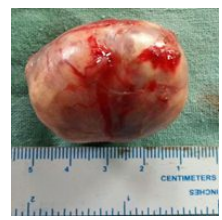

figure3]

The post operative histopathological report was of neurilemmoma.Our patient had an uneventful postoperative recovery. Total excision resulted in complete resolution of symptoms with no recurrence or paraesthesia till date.

Discussion:-

Schwannoma, first described in 1908 by Verocay, commonly occurs between the age of 30 and 50 years old. Neurilemmoma/schwannoma is a benign tumor which arises from and consists solely of Schwann cells [1]. Between 25% to 45% of all neurilemmomas occur in the head and neck region, and 20% of these lesions occur in the intraoral area.

Intraoral neurilemmomas arise mainly from the tongue, followed by the palate, floor of mouth, buccal mucosa, gingiva, lips, and vestibular mucosa [6,7]. Neurilemmomas usually occur in the second or third decade of life, but can develop at any age. There is no definite sex predilection[8]. Masses are typically between 0.5 to 3 cm in size, rarely exceeding 5 cm[6,9].

Minor salivary gland tumors are rare, comprising 0.5% to 1% of all epithelial salivary gland tumors.Neurilemmoma in floor of mouth must be differentiated from minor salivary gland tumors. Since 80% to 90% of all minor salivary gland tumors show malignancy[10]. The imaging features of low-grade malignant salivary gland lesions resemble those of benign salivary gland tumors[11]. Therefore, when dealing with sublingual area masses, clinicians should keep in mind the possibility of malignancy and sublingual gland tumor, though definitive diagnosis depends on histopathologic examination. So the dilema faced by the clinician is to go for pre operative biopsy or not.

Salivary gland tumors include a wide range of tumor types, which can be either benign or malignant. Approximately 85 percent arise in the parotid; the remaining originate in the submandibular, sublingual, and minor salivary glands, which are located throughout the submucosa of the mouth and upper aerodigestive tract [12]. Approximately 25 percent of parotid tumors are malignant, compared with 40 to 45 percent of submandibular gland tumors, 70 to 90 percent of sublingual gland tumors, and 50 to 75 percent of minor salivary gland tumors.

Schwannomas are usually solitary lesions; however, some are seen as multiple lesions as part of neurofibromatosis type [1] .

Clinical evidence of the tumor usually does not present for a long time. The most common symptom is a slow-growing mass . Neurological symptoms and pain are rare. In schwannomas malignant transformation is very rare .

Diagnostic investigations include computed tomography (CT), magnetic resonance imaging (MRI), ultrasound scan and FNA.

MRI is the best choice in detecting the extent of the tumor and correlates well with the operative findings. In T1-weighted MR images, the signals of the lesions are isointense to that of muscle, and hyperintense to that of muscle on T2-weighted images[13]. Schwannomas have specific MRI properties, including specific signs (split-fat sign, fascicular sign, target sign) and signal patterns (that is, isointense T1 signal relative to skeletal muscle; increased and slightly heterogeneous T2 signal)

Neurilemmomas show well-defined boundaries and appear homogeneous on CT images. When CT images show heterogeneous lesions, malignant changes in the neurogenic tumor may be suspected [14]

Biswas et al. have reported their 10 years of experience regarding extracranial head and neck schwannomas, and in their report, only 6 percent of patients could have been diagnosed preoperatively on the basis of clinical findings, CT and MRI scans, and FNAC[15] .

Microscopic evaluation of neurilemmoma shows two main patterns. The Antoni A pattern consists of densely-packed spindle cells. These cells are arranged in a typical, palisading figure surrounding acellular, eosinophilic areas known as Verocay bodies. The Antoni B pattern, the loose hypocellular arrangement, lies beside the Antoni A pattern. Positive staining for the neural crest marker, S-100 protein, is an important characteristic for diagnosis[9,16,8]. The tumor in our patient showed characteristics of typical neurilemmoma.

The treatment of schwannomas is problematic. Because of resistance to radiotherapy, surgical excision is necessary for optimal treatment [7]. The current treatment modality for neurilemmoma is complete surgical excision[2]. The intraoral approach avoids extraoral scarring and is preferred. However, in some cases, intraoral excision may not be adequate for complete removal of the mass. Incomplete surgical removal of a neurilemmoma may result in recurrence[17]. Transhyoid excision can be a cosmetic alternative. The choice of surgical approach is based on tumor size and location[18].

Kang et al. reported that more than half of the surgically treated cases exhibited postoperative neural deficits that were primarily caused by iatrogenic injury to either the nerve of origin or adjacent neural ending . In our case, no neural deficits or other problems presented after total excision of the tumor[19].

The malignant potential of extracranial schwannomas and risk of recurrence after surgical resection are unclear , while in most studies investigating extracranial schwannomas, recurrence or malignant transformation of the tumor have not been reported [8] . During one-year follow-up of the present case, there was no evidence of recurrence and prognosis was excellent.

Conclusion:-

Schwannoma of the salivary gland is a particularly rare form of an extracranial neurogenic tumor. Our findings indicate good prognosis in an unusual case of a sublingual schwannoma in a 55-year old female treated by surgical excision with no recurrence within 7 months of follow-up.

REFERENCES:

1. Arda HN, Akdogan O, Arda N, Sarikaya Y. An unusual site for an intraoral schwannoma: a case report. Am J Otolaryngol. 2003;24:348–350.[PubMed]
2. Naik SM, Goutham MK, Ravishankara S, Appaji MK. Sublingual Schwannoma: a rare clinical entity reported in a hypothyroid female. Int J Head Neck Surg. 2012;3:33–39 .
3. Weiss SW, Goldblum JR. Benign tumors of peripheral nerves. In: Strauss M, editor. Enzinger and Weiss's soft tissue tumors. 4th ed. St. Louis: Mosby; 2001. pp. 11112.–207
4. Golfarb A Pathologist Dept of anatomical and clinical pathology , South West Hospital Cortez,CO. American journal of otolarinngology-head and neck medicine and surgery. 29:119-122, Sawhney and Carron MA Mathog RH(2008)
5 Yahya Sulaiman Marhoon Al-Alawi,Arif Ali,PAM Saparamadu,Yahya Al Badaai;Oman Medical Journal2013 vol 29,no.2 sublingual gland schwannoma:a rare case at unusual site.
6. Tsushima F, Sawai T, Kayamori K, Okada N, Omura K. Schwannoma in the floor of the mouth: a case report and clinicopathological studies of 10 cases in the oral region. J Oral Maxillofac Surg Med Pathol. 2012;24:175–179.
7. Gallo WJ, Moss M, Shapiro DN, Gaul JV. Neurilemoma: review of the literature and report of five cases. J Oral Surg. 1977;35:235–236.[PubMed]
8. Neville BW, Damm DD, Allen CM, Bouquot JE. Oral and maxillofacial pathology. 2nd ed. Philadelphia: W.B. Saunders; 2002. pp. 456–457.
9. de Bree R, Westerveld GJ, Smeele LE. Submandibular approach for excision of a large schwannoma in the base of the tongue. Eur Arch Otorhinolaryngol. 2000;257:283–286.[PubMed]
10. Rinaldo A, Shaha AR, Pellitteri PK, Bradley PJ, Ferlito A. Management of malignant sublingual salivary gland tumors. Oral Oncol. 2004;40:2–5.[PubMed]
11. Lee YY, Wong KT, King AD, Ahuja AT. Imaging of salivary gland tumours. Eur J Radiol. 2008;66:419–436.[PubMed
12 Guzzo M, Locati LD, Prott FJ, et al. Major and minor salivary gland tumors. Crit Rev Oncol Hematol 2010; 74:134
13. La'porte SJ, Juttla JK, Lingam RK. Imaging the floor of the mouth and the sublingual space. Radiographics. 2011;31:1215–1230.[PubMed]
14. Cohen LM, Schwartz AM, Rockoff SD. Benign schwannomas: pathologic basis for CT inhomogeneities. AJR Am J Roentgenol. 1986;147:141–143.[PubMed]
15 Biswas D, Marnane CN, Mal R, Baldwin D. Extracranial head and neck schwannomas - a 10-year review. Auris Nasus Larynx. 2007;34:353–359. doi: 10.1016/j.anl.2007.01.006.
16. Laviv A, Faquin WC, August M. Schwannoma (neurilemmoma) in the floor of the mouth: Presentation of two cases. J Oral Maxillofac Surg Med Pathol. 2015;27:199–203.
17. AL-Alawi YSM, Kolethekkat AA, Saparamadu PAM, Al Badaai Y. Sublingual gland schwannoma: a rare case at an unusual site. Oman Med J. 2014;29 doi: 10.5001/omj.2014.41. [Cross Ref]
18. Hsu YC, Hwang CF, Hsu RF, Kuo FY, Chien CY. Schwannoma (neurilemmoma) of the tongue. Acta Otolaryngol. 2006;126:861–865.[PubMed
19 Kang GC, Soo KC, Lim DT. Extracranial non-vestibular head and neck schwannomas: a ten-year experience. Ann Acad Med. 2007;36:233–238.