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General Surgery

HYPOGLOSSAL NEURILEMMOMA IN THE SUBMANDIBULAR REGION A DIAGNOSTIC DILEMMA

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ABSTRACT Pleomorphic adenoma is a common benign salivary gland tumor, predominantly found in the parotid gland, with only 10% occurring in the submandibular gland. It is a mixed tumor comprising epithelial, myoepithelial, and mesenchymal components. Hypoglossal schwannomas, on the other hand, are extremely rare, with an undefined incidence. These tumors arise from Schwann cells, responsible for myelin sheath formation, and can occur anywhere along the course of the hypoglossal nerve. Due to their proximity to the submandibular gland, they are frequently misdiagnosed as submandibular pleomorphic adenomas. We report a 24-year-old female presented with a gradually enlarging right submandibular swelling over two years, with occasional swallowing discomfort. Clinical evaluation revealed a firm, spherical 6×6 cm mass without speech abnormalities. Ultrasonography and CECT neck suggested pleomorphic adenoma, confirmed by FNAC. The patient underwent elective submandibular gland excision. Intraoperatively, a separate lesion arising from the hypoglossal nerve was discovered and carefully dissected. Postoperatively, she had mild tongue deviation and facial asymmetry, which resolved in three weeks. Histopathology confirmed a benign schwannoma. Preoperative diagnosis is challenging, with studies showing only 6% accuracy. Similar cases have been misdiagnosed as submandibular adenomas, emphasizing the diagnostic dilemma. MRI features, such as split-fat, fascicular, and target signs, aid diagnosis, but findings are not always conclusive. Histopathological examination remains the gold standard. Thus Hypoglossal neurilemmoma is a rare entity often mistaken for submandibular pleomorphic adenoma. Awareness of this differential diagnosis is crucial to prevent misdiagnosis and ensure appropriate surgical management.

KEYWORDS : Hypoglossal Neurilemmoma in the Submandibular region a diagnostic dilemma

INTRODUCTION

Pleomorphic adenoma is a salivary gland tumor and mostly found in the parotid gland. A pleomorphic tumor is a mixed tumor (benign mixed tumor) consisted of epithelium, myoepithelial, mesenchyme and made of a view component variation of it.

Almost 85% of benign epithelial salivary gland tumors arise from the parotid gland and only 10% in the submandibular gland. Pleomorphic adenoma typically present as a slow growing, unilateral and painless mass of salivary glands, with a predilection for recurrence and risk of malignant transformation (about 1.5% up to 5 years and increases to 9.5% after more than 15 years (1) The submandibular glands are paired major salivary glands that lie in the submandibular triangle. The glands have a superficial and deep lobe separated by the mylohyoid muscle (2) Schwann cells, or neurilemma, are cells that are responsible for the production of the myelin sheath around neuronal axons. The vast majority of tumors of the peripheral nervous system arise from the cells of Schwann, rather than from the nerve cells themselves. Most of these tumors are benign and designated as "schwannomas" or "neurilemommas.(3) Schwannomas of the hypoglossal nerve are extremely rare with incidence clearly not defined. Hypoglossal schwannomas can arise anywhere along the course of the nerve and are commonly reported to affect middle-aged females. The Hypoglossal Nerve ie the 12th Cranial Nerve, is mainly an efferent nerve for the tongue musculature. The nerve originates from the medulla and travels caudally and dorsally to the tongue. The hypoglossal nerve is mainly a somatic efferent (motor) nerve to innervate the tongue musculature. The nerve also contains some sympathetic postganglionic fibers from the cervical ganglia, which innervates tongue vessels and some small glands in the oral mucosa.(4)

The hypoglossal nerve lies in the bed of the deep lobe of the submandibular gland lying in close relation to the submandibular gland. Kaye et al. proposed a classification system based on the location of the schwannoma as intracranial (type A), intracranial/ extracranial (type B), or extracranial (type C). The intracranial/ extracranial tumors (type B), which extend through and expand the hypoglossal canal, are often described as “dumb-bell shaped” based on their appearance on imaging studies and are the most common type.

CASE REPORT

A 24 year old female presented with a swelling in the right submandibular region which was insidious in onset and progressed in size gradually over course of 2 years. The patient was asymptomatic at presentation with occasional discomfort while swallowing. The patient predominantly presented to the hospital due to cosmetic reasons.

A through clinical evaluation revealed a firm spherical swelling in the right submandibular region with of about 6*6 cm, with no speech abnormalities or heaviness of the tongue.

As a initial evaluation a USG neck was performed which revealed a well defined hypoechoic round lesion with smooth margin in the right submandibular gland, likely suggestive of pleomorphic adenoma.

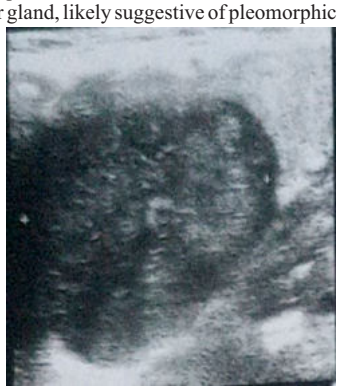

Figure- 1- Ultrasonography -Well defined hypoechoic lesion with smooth margins in submandibular region S/o submandibular adenoma

The patient was further evaluated with a CECT neck and Fine needle aspiration cytology.

The CT neck revealed- a well defined mild heterogeneously enhancing lesion in the right submandibular region, likely representing pleomorphic adenoma. The diagnosis was confirmed with a FNAC where the smear showed benign ductal epithelial cells, in fibomyxoid background, which were suggestive of pleomorphic adenoma.

The patient was the planned for a elective submandibular gland excision.

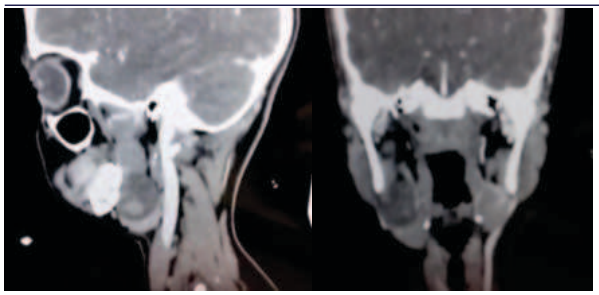

Figure 2- Well defined heterogeneously enhancing lesion in the submandibular region s/o Pleomorphic adenoma

A infra-mandibular linear incision was taken and the submandibular gland was isolated. The gland appeared bulky and was excised.

However on excision of gland , a bulge over the posterior inferior aspect of the gland was noted. On further exploration a well defined ovoid lesion noted arising from the hypoglossal nerve.

Figure 3– solitary lesion arising from the hypoglossal nerve

The lesion appeared to be arising from the nerve sheath and was carefully dissected off, keeping the nerve intact. The nerve was further traced proximally to confirm its anatomy.

The patient had an uneventful post operative course. Post operatively she mild drooping of angle of mouth with deviation of tongue on protrusion to the opposite side, which resolved gradually over 3 weeks. The histo-pathological analysis revealed a benign capsulated neoplasm comprising of cellular and hypocellular areas. The cellular areas show spindle cells showing mild nuclear pleomorphism, occasionally showing palisading. Suggestive of neurilemmoma. Schwannomas can develop from the glossopharyngeal, accessory, and hypoglossal nerves among the cranial nerves, with acoustic neuroma being the most common type, originating from the vestibulocochlear nerve

DISCUSSION

Neurilemmoma, also known as schwannoma, is a noncancerous tumor that originates from Schwann cells and is composed entirely of them. Around 25% to 45% of extracranial schwannomas occur in the head and neck region(5)

Biswas et al. documented their ten-year experience with extracranial head and neck schwannomas. According to their report, only 6% of patients were accurately diagnosed before surgery based on clinical evaluation, CT and MRI imaging, and fine-needle aspiration. These findings were consistant with our case in which even after extensive preoperative testing, the schwannoma could not be detected(6)

Similarly, Ranjan et al, reported a case with comparable findings, initially diagnosed clinically as a submandibular adenoma, but later identified as a schwannoma through further FNA evaluation(7) these findings were consistent with our case. A similar case report was also reported by Aslan et al, although the authors could not specify the exact origin(5) Thus a neurilimomma in the submandibular region remains a diagnostic dilemma.

While schwannomas display characteristic MRI features, including specific signs (split-fat sign, fascicular sign, and target sign) and distinct signal patterns (isointense T1 signal relative to skeletal muscle with an increased, slightly heterogeneous T2 signal)(8), these findings are not universally picked up as seen in this case and a definitive diagnosis can only be confirmed through histopathological examination.

CONCLUSION-

Hypoglossal neurilemmoma is a rare condition with an unclear incidence. Because of its close proximity to the submandibular gland, it is often mistaken for submandibular pleomorphic adenoma. Therefore, hypoglossal neurilemmoma should always be considered as a differential diagnosis in cases of submandibular adenoma.

REFERENCES

1. Farhat F, Asnir RA, Yudhistira A, Daulay ER, Sagala IP. An Uncommon Occurrence of Pleomorphic Adenoma in the Submandibular Salivary Gland: A Case Report. Open Access Maced J Med Sci. 2018 Jun 17;6(6):1101–3.
2. Grewal JS, Jamal Z, Ryan J. Anatomy, Head and Neck, Submandibular Gland. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2023 [cited 2024 Jan 12]. Available from: http://www.ncbi.nlm.nih.gov/books/NBK542272/
3. Davis DD, Kane SM. Neurilemmoma. In: StatPearls [Internet] [Internet]. StatPearls Publishing; 2023 [cited 2024 Jan 12]. Available from: https://www.ncbi.nlm.nih.gov/books/NBK560827/
4. Kim SY, Naqvi IA. Neuroanatomy, Cranial Nerve 12 (Hypoglossal). In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2023 [cited 2024 Jan 12]. Available from: http://www.ncbi.nlm.nih.gov/books/NBK532869/
5. Aslan G, Cinar F, Cabuk FK. Schwannoma of the submandibular gland: a case report. J Med Case Reports. 2014 Jun 26;8:231.
6. Biswas D, Marnane CN, Mal R, Baldwin D. Extracranial head and neck schwannomas--a 10-year review. Auris Nasus Larynx. 2007 Sep;34(3):353–9.
7. Ranjan SK, Sinha M, Sharan A, Munda VS, Usha P, Yadav SK. Schwannoma of the Submandibular Gland: A Rare Case Report. J Clin Diagn Res JCDR. 2016 Feb 1;10(2):PD23.
8. Beaman FD, Kransdorf MJ, Menke DM. Schwannoma: radiologic-pathologic correlation. Radiogr Rev Publ Radiol Soc N Am Inc. 2004;24(5):1477–81.