DOI: 10.4274/MMJ.galenos.2024.37729
Medeni Med J 2024;39:132-135

Case Report

Solitary Submandibular Schwannoma Mimicking a Salivary Gland Tumor in a Child

Bir Çocukta Tükürük Bezi Tümörünü Taklit Eden Soliter Submandibular Schwannoma

Kim Yew Richard WONG[1], Ikram HAKIM[1], Halimuddin SAWALI[2], Rachel Cui Ai LIM[1], Nur Kamilia MOHD MOHSIN[3]

[1]Hospital Tawau, Department of Otorhinolaryngology-Head and Neck Surgery, Sabah, Malaysia
[2]Hospital Queen Elizabeth, Department of Otorhinolaryngology-Head and Neck Surgery, Sabah, Malaysia
[3]Hospital Queen Elizabeth, Department of Pathology, Sabah, Malaysia

ABSTRACT

Tumors occurring in the submandibular space are infrequent among pediatric patients, and benign peripheral nerve tumors in this region are exceptionally rare. This study describes the uncommon occurrence of a schwannoma in the submandibular space in a child and describes its management. A 7-year-old child presented with a gradually enlarging swelling over a 7-month period in the submandibular region, clinically resembling a salivary gland tumor. There were no associated marginal mandibular, lingual, or hypoglossal nerve palsy. The mass was excised completely, and histopathological examination revealed it to be a schwannoma. It is appropriate to consider benign peripheral nerve tumors, such as schwannoma, in the differential diagnosis of submandibular space tumors in children.

Keywords: Submandibular gland neoplasms, neurilemmoma, salivary gland neoplasms

ÖZ

Submandibular boşlukta meydana gelen tümörler pediatrik hastalar arasında sık görülmez ve bu bölgedeki benign periferik sinir tümörleri son derece nadirdir. Bu çalışmada, bir çocukta submandibular alanda nadir görülen bir schwannoma olgusu ve tedavisi anlatılmaktadır. Yedi yaşında bir çocuk, submandibular bölgede 7 aylık bir süre içinde giderek büyüyen ve klinik olarak tükürük bezi tümörüne benzeyen bir şişlik ile başvurdu. Kitle ile ilişkili marjinal mandibular, lingual veya hipoglossal sinir felci yoktu. Kitle tamamen eksize edildi ve histopatolojik incelemede schwannoma olduğu görüldü. Çocuklarda submandibular boşluk tümörlerinin ayırıcı tanısında schwannoma gibi benign periferik sinir tümörlerinin düşünülmesi gerekmektedir.

Anahtar kelimeler: Submandibular bez neoplazmları, nörilemmoma, tükürük bezi neoplazmları

INTRODUCTION

Salivary masses in children comprise 10% of all tumors affecting the head and neck in the pediatric population. While uncommon, it is about five times more likely to be malignant than in an adult, with rates of 50% and 10%, respectively. The parotid gland (66%) emerged as the most frequently affected site, followed by the submandibular gland (34%). Benign lesions accounted for 94% of submandibular gland tumors. Pleomorphic adenoma was the predominant benign tumor (92%), whereas acinic cell carcinoma stood out as the leading malignancy (64%)[1]. Other benign tumors of the submandibular space include Warthin's tumor, embryoma, monomorphic adenoma, lipoma, and teratoma, listed in decreasing order of occurrence[2].

CASE REPORT

A 7-year-old girl of Bajau descent, with no medical comorbidities, presented to us with a history of painless, slow-growing right submandibular swelling for 7 months. There were no associated skin changes or pus discharge. There were no history of preceding trauma or insect bite. On examination, there was a right submandibular swelling measuring 4x3 cm, which was firm in consistency

Address for Correspondence: K.Y.R. Wong, Hospital Tawau, Clinic of Otorhinolaryngology-Head and Neck Surgery, Sabah, Malaysia
E-mail: richardwky@gmail.com **ORCID ID:** orcid.org/0000-0002-0165-0895

Received: 26 February 2024
Accepted: 02 April 2024
Online First: 15 April 2024

Cite as: Wong KYR, Hakim I, Sawali H, Lim RCA, Mohd Mohsin NK. Solitary Submandibular Schwannoma Mimicking a Salivary Gland Tumor in a Child. Medeni Med J 2024;39:132-135

and non-tender (Figure 1). No punctum or skin changes were noted. There were no remarkable findings in the oral cavity and no associated marginal mandibular, lingual, or hypoglossal nerve palsy. There was no medialization of the lateral pharyngeal wall on flexible nasopharyngolaryngoscopy, with normal supraglottic and glottic structures observed. On the basis of our initial clinical assessment, a provisional diagnosis of a submandibular gland pleomorphic adenoma was made.

Tru-cut needle biopsy of the right submandibular swelling, however; revealed a low-grade spindle cell lesion, with features favoring a benign peripheral nerve sheath tumor.

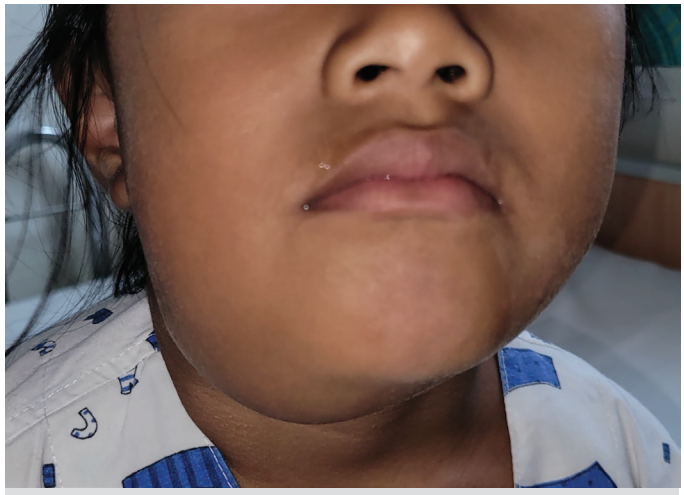
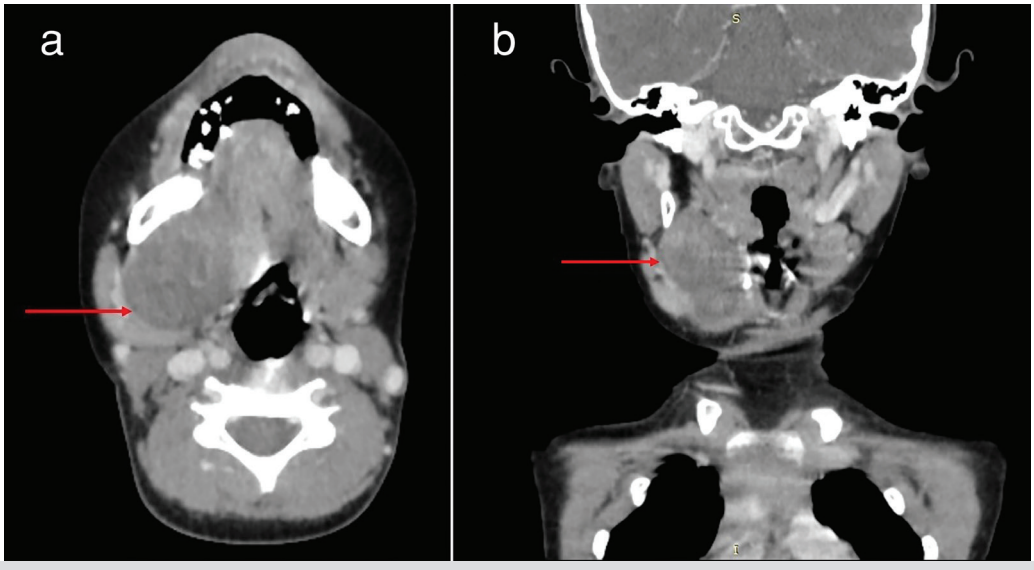


Figure 1. Right submandibular swelling measuring 4x3 cm.

Computed tomography (CT) scan of the neck (Figure 2) showed a large, well-defined, heterogeneously enhancing hypodense mass (45 Hounsfield unit) at the right submandibular space measuring approximately 4.7x3.2x3.5 cm (AP x W x CC). The mass displaces the right submandibular gland posteriorly and extends from the level of the right medial pterygoid muscle superiorly to the level of the hyoid bone inferiorly. No significant cervical lymphadenopathy was observed.

The patient underwent excision of the right submandibular mass under general anesthesia. A skin incision was made 2 cm below the angle of the mandible, and the subplatysmal flap was raised. The marginal mandibular nerve was protected using the Hayes-Martin maneuver. Upon entering the submandibular space, there was a solid whitish mass with a smooth surface measuring 3x5 cm (Figure 3). The lingual nerve and submandibular gland were identified and preserved. The originating nerve of the tumor was not determined in this case. Nonetheless, a meticulous intracapsular dissection was performed to completely excise the mass.

Histopathological examination of the specimen confirmed that it was a benign peripheral nerve sheath tumor, which was consistent with Schwannoma; as sections show a thinly encapsulated lesion composed of proliferation of spindle cells arranged in hypercellular (Antoni A) and hypocellular areas (Antoni B) (Figure 4). Verocay bodies were observed in the hypercellular areas (Figure 5). This tumor was further classified as a conventional schwannoma because it does not exhibit features specific to other special subtypes; for example,



Figures 2. Contrast enhanced computed tomography scan of the neck in axial (a) & coronal (b) views showing the right submandibular mass (red arrow).

the ancient, cellular, plexiform, or microcystic subtypes. The child recovered well postoperatively with no recurrence or nerve deficit within 6 months of follow-up. The patients parents provided written informed consent for the publication of this article.

DISCUSSION

Schwannoma, initially described by Verocay in 1908, typically manifests in individuals aged between 30 and 50 years. They are benign tumors that are solitary and well differentiated and originate from Schwann cells. Approximately 25-45% of extracranial schwannomas are found in the head and neck region; with the temporal bone, lateral neck, and paranasal sinuses being the most frequently affected areas. Schwannomas typically occur as solitary lesions; however, in some cases, they may manifest as multiple lesions as part of neurofibromatosis type 2. The nerve of origin is unidentifiable in approximately 10-40% of schwannomas[3].

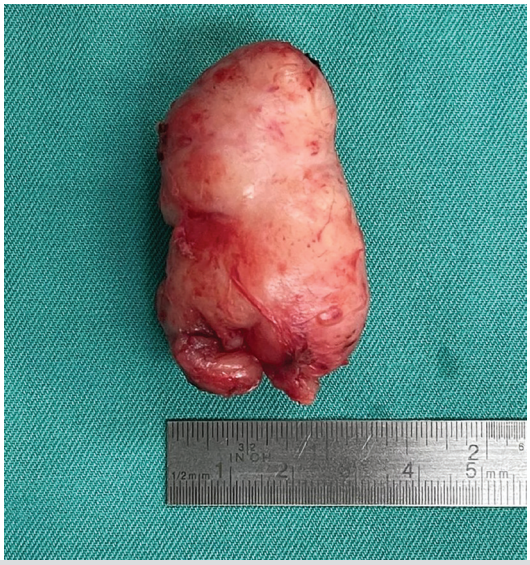


Figure 3. Operative specimen showing a solid whitish mass with smooth surface which was completely excised.

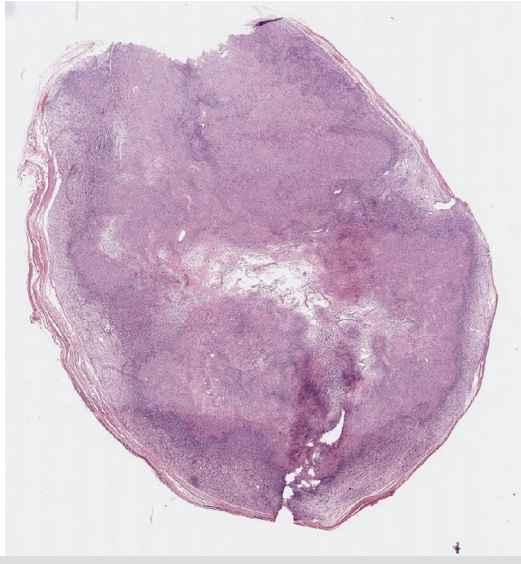


Figure 4. Histopathologic examination of the surgical specimen demonstrated a thinly encapsulated lesion composed of proliferation of spindle cells arranged in hypercellular (Antoni B) and hypocellular areas (Antoni B).

Extracranial schwannomas situated in the salivary glands are infrequent, with the majority arising in the parotid gland from a peripheral branch of the facial nerve. Schwannomas originating in the submandibular space are exceedingly rare, and only a limited number of cases have been reported. Patients present with a painless and mobile mass in the submandibular region. These benign tumors tend to displace rather than infiltrate the associated nerve, which accounts for the absence of nerve palsy[4].

Ultrasonography reveals a well-defined solid hypoechoic mass, not unlike pleomorphic adenomas. CT characteristics of schwannomas include well-defined tumors with low or soft tissue attenuation and either homogeneous or heterogeneous enhancement. Magnetic resonance imaging (MRI) is recognized as the most effective imaging technique for diagnosing schwannomas. On MRI, schwannomas display low signal intensity on T1-weighted images and high signal intensity on T2-weighted images. In T2-weighted images, a distinctive target signal pattern is evident, featuring heightened peripheral signal intensity and reduced central signal intensity[5].

Schwannoma exhibits a distinctive histological profile characterized by an encapsulated lesion originating from a nerve. It is composed of a close intermingling of spindle

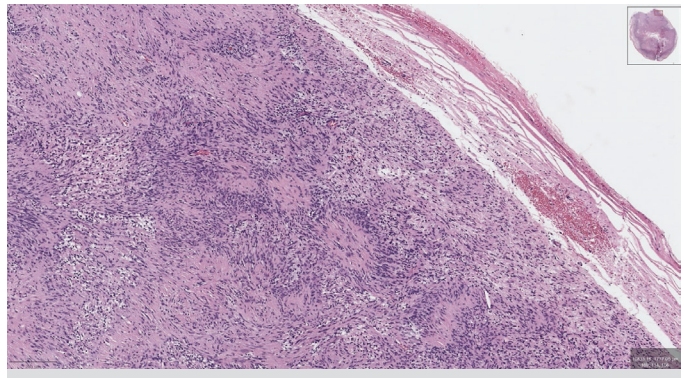


Figure 5. Prominent nuclear palisading forming Verocay bodies are seen in the hypercellular areas.

cells that form highly cellular areas known as Antoni type A, along with less cellular and myxoid Antoni type B areas. Within the palisades of Antoni type A cells, there are regions without nuclei referred to as Verocay bodies[6].

Schwannomas found in various regions of the human body have demonstrated immunoreactivity to proteins S-100 and vimentin. Furthermore, varying immunoreactivity to EMA and GFAP was identified in areas corresponding to the perineurium. Although uncommon, positivity for additional markers like CD56 and CD57 has been noted in both benign and malignant schwannomas[7].

The preferred approach for treating schwannoma typically involves removing the tumor while preserving the associated nerve[8]. The usual challenge lies in the inability to identify the nerve of origin, and even with nerve-preserving intracapsular enucleation, preserved nerve function post-surgery is not guaranteed. In some cases, postoperative neurological deficits may help establish the tumor's origin[9].

After a thorough surgical excision, there is no need for long-term follow-up; as recurrence is rare after surgery. However, in cases of incomplete excision, it is advisable to undergo annual ultrasonography and review[10].

In conclusion, submandibular space schwannoma is a rare entity; and this, to the best of our knowledge; is the first reported case in the pediatric age group. Diagnosis of the lesion is typically achieved through a contrast-enhanced CT scan of the neck coupled with fine needle aspiration cytology. Complete surgical resection is generally considered curative.

Acknowledgments: We would like to thank the Director General of Ministry of Health Malaysia for his permission to publish this article as a case report.

Ethics

Informed Consent: Patient's guardian's written informed consent was obtained.

Author Contributions

Surgical and Medical Practices: K.Y.R.W., I.H., H.S., R.C.A.L., N.K.M.M., Concept: K.Y.R.W., I.H., H.S., R.C.A.L., N.K.M.M., Design: K.Y.R.W., I.H., H.S., R.C.A.L., N.K.M.M., Data Collection and/or Processing: K.Y.R.W., I.H., R.C.A.L., N.K.M.M., Analysis and/or Interpretation: K.Y.R.W., N.K.M.M., Literature Search: K.Y.R.W., Writing: K.Y.R.W., I.H., H.S.

Conflict of Interest: The authors have no conflict of interest to declare.

Financial Disclosure: The authors declared that this study has received no financial support.

REFERENCES

1. Jesberg P, Monzon A, Gitomer SA, Herrmann BW. Pediatric primary salivary gland tumors. Am J Otolaryngol. 2023;44:103948.
2. Lesperance Marci M, Flint PW. Salivary gland disease in children. In: Cummings Pediatric Otolaryngology. 1st ed. Elsevier; 2015. p. 304-5.
3. 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. 2016;10:PD23-4.
4. Bondy PC, Block RM, Green J. Ancient schwannoma of the submandibular gland: A case report. Ear Nose Throat J. 1996;75:781-3.
5. Park KW, Lee DH, Lee JK, Lim SC. A clinical study of submandibular schwannoma. Oral Surg Oral Med Oral Pathol Oral Radiol. 2022;133:e6-9.
6. Bansal V, Aggarwal P, Wadhwan V, Bansal A, Kapoor S, Yadav M. Giant solitary schwannoma of submandibular salivary gland-a rare entity. J Maxillofac Oral Surg. 2017;16:382-6.
7. dos Santos JN, Silva Gurgel CA, Gonçalves Ramos EA, Pereira Júnior FB, Crusoé-Rebello IM, Oliveira MC. Plexiform schwannoma mimicking a salivary gland tumor: An unusual case report diagnosed in pediatric patient. Int J Pediatr Otorhinolaryngol Extra. 2011;6:317-21.
8. Kang GC, Soo KC, Lim DT. Extracranial non-vestibular head and neck schwannomas: A ten-year experience. Ann Acad Med Singap. 2007;36:233-40.
9. Loterh SS, Prabhu V, O'Brien C. Submandibular schwannoma: Is excision a 'gold standard treatment'? BMJ Case Rep. 2015;2015:bcr2015209900.
10. Patel M, Srinivasan B, Sharma S. The diagnostic dilemma of a parasympathetic schwannoma in the submandibular gland. Ann R Coll Surg Engl. 2015;97:e13-4.