Primary Isolated Solitary Schwannoma of the Uvula

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Schwannomas of the soft palate are rare, and a solitary, isolated schwannoma originating from or involving the uvula has never been reported before.

A 35-year-old woman presented with complaints of “something stuck during swallowing” for the last 6 months. She noted a painless mass hanging from her soft palate on looking in the mirror. Examination revealed a smooth, mucosa-covered globular mass hanging from the posterior midline soft palate, replacing the uvula, partly extending to the adjacent soft tissue (Figure 1). On gentle probing, it appeared firm, insensitive, nontender, intimately attached with the lower two-thirds of uvula. The posterior pharyngeal wall, rest of the oropharynx and oral cavity were unremarkable. There were no signs of local and systemic inflammation. The patient had no immunocompromise and comorbidities and was otherwise healthy. She had no history of recent febrile illness, acute coryza with sore throat, or intake of drug/food that could lead to allergic manifestations. An excision biopsy of the lesion under general anesthesia was planned.

During surgery, the mass was found to originate from the uvula and encroached to the adjacent soft palate. It was completely removed, preserving the upper third of uvula. Grossly, the mass was oblong, smooth, fleshy, measuring approximately $4 \times 1.5 \times 2$ cm, with prominent superficial vasculature (Figure 2). Histopathology showed dense spindle cell population in wavy bundles. There were more cellular areas (Antoni A) with palisading of spindle and round cell nuclei (Verocay bodies), often with degenerative atypia, along with less cellular areas (Antoni B) containing edematous stroma where fibers and cells formed no distinctive pattern (Figure 3). There was no appreciable capsule, and no evidence of malignancy. The features strongly suggested a schwannoma. The lesion was diffusely positive for S-100 protein, confirming the diagnosis. At 6 months follow-up, the patient did not experience recurrence or velopharyngeal insufficiency.

Schwannomas (aka neurinoma, neurilemmoma) are benign primitive neuroectodermal nerve sheath tumors originating from differentiated Schwann cells responsible for neural myelination. Histologically, they differ from neurofibromas by the presence of distinct capsule, homogeneous cell types of Schwann cell origin, distinctive Verocay bodies (Antoni A areas), and dilated vessels with surrounding hyalinization. Schwannomas are relatively frequent in the head-neck, constituting 25% to 40% of the overall incidence.1 However, intraoral schwannomas are essentially uncommon (1%-12%), mostly involving the anterior tongue and buccal floor.1 Soft palate is only rarely involved.2 There are reports of indirect involvement of soft palate and uvula in schwannomas presenting as large parapharyngeal tumors.3,4 However, extensive PubMed/MEDLINE search with keywords “soft palate,” “uvula,” and “schwannoma” revealed no published records of isolated, solitary schwannoma originating primarily from and/or involving the uvula. The latest systematic literature review on 46 patients exclusively with palatal schwannoma documented since 1985 revealed soft palate involvement in less than a third, with only one patient having midline posterior soft palate involvement that, however, spared the uvula.2

Essentially therefore, primary, isolated, solitary schwannoma is the diagnosis of exclusion for an uvular mass. Common differential diagnoses would include solid germ cell lesions (epidermoid cyst, dermoid, teratoma), mucocele, lymphoepithelial cysts, and salivary gland tumors (including pleomorphic adenoma).5 Possibilities of uvular swelling due to a parapharyngeal neoplasm,6 or in response to allergy (angioneurotic edema, acute coryza), should also be considered.

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considered. However, since fine needle aspiration cytology from an uvular mass is mostly nonconfirmatory, and due to the technical difficulties associated it, it is generally impossible to reach a preoperative diagnosis, adding to the clinical dilemma. In our patient, despite a thorough history and clinical examination, the diagnosis of schwannoma was least suspected. Complete excision is the treatment of choice, which might also be the only mean for proper diagnosis through histopathology.

The uvular lesion described here partly deviated from the classic clinicohistologic description of schwannoma. The tumor was poorly encapsulated and submucosal, but without surface ulceration and pedunculation, the 2 features commonly associated with this variant. Also, central schwannomas generally originate from bone or indirectly result in bone erosion, contrary to the peripheral types that have a soft tissue predilection. However, soft palate schwannomas seldom involve bone; instead, the neoplasm in our patient was found intimately associated with the uvula. Nevertheless, the isolated uvular mass described here had telltale histologic features of schwannoma like Antoni A (with Verocay bodies) and B areas, prominent vessels with perivascular hyalinization, and spindle cells with degenerative nuclear atypia.
Schwannoma as a solitary, isolated uvular neoplasm makes our observation worth presenting. We believe schwannoma should be considered a possibility when dealing with a submucosal, well-circumscribed but poorly encapsulated soft tissue lesion in the soft palate and uvula.

Authors’ Note
Informed consent in writing has been obtained from the patient prior to submission of the report.

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References
1. Gainza-Cirauqui ML, Eguía-Del Valle A, Martínez-Conde R, Coca-Meneses JC, Aguirre-Urizar JM. Ancient schwannoma of the hard palate. An uncommon case report and review. J Clin Exp Dent. 2013;5(1):e62-65.
2. Dokania V, Rajguru A, Mayashankar V, Mukherjee I, Jaipuria B, Shere D. Palatal schwannoma: an analysis of 45 literature reports and of an illustrative case. Int Arch Otorhinolaryngol. 2019;23(3):e360-370.
3. Ramdass AA, Yao M, Natarajan S, Bakshi PK. A rare case of vagus nerve schwannoma presenting as a neck mass. Am J Case Rep. 2017;18:908-911.
4. Bhardwaj M, Saini S, Kaur K, Choudhary S, Hooda S. Anaesthetic management of a case of schwannoma with intraoral extension. Egypt J Anaesth. 2015;31(2):215-217.
5. Daram S, Ulualp SO, Uddin N. Epidermoid cyst of the uvula in a child. SAGE Open Med Case Rep. 2016;4:1-3.
6. Wareing MJ, Irving RM, Moffat DA. Parapharyngeal space tumour presenting as recurrent uvular oedema. J Laryngol Otol. 1993;107(7):640-641.
7. Suga K, Muramatsu K, Uchiyama T, Takano N, Shibahara T. Congenital epidermoid cyst arising in soft palate near uvula: a case report. Bull Tokyo Dent Coll. 2010;51(4):207-211.
8. Hribernik SJ, Gould AR, Alpert B, Jones JL. Well-circumscribed mass of the lateral floor of the mouth. J Oral Maxillofac Surg. 1992;50(7):741-746.