IOSR Journal of Dental and Medical Sciences (IOSR-JDMS)
e-ISSN: 2279-0853, p-ISSN: 2279-0861.Volume 22, Issue 3 Ser.6 (March. 2023), PP 36-41
www.iosrjournals.org

Cervical cystic schwannoma of the vagus nerve Case report and review of literature.

Kamel Bouaita; Lynda Atroune; Soumia Benallag, t.selmane
Cherchell hospital

Abstract
Introduction: Cervical schwannomas are benign peripheral nerve tumors developed exclusively from Schwann cells. Involvement of the cervical vagus nerve is relatively rare; the treatment of these tumors is surgical.
Case report: A 26-year-old patient consulted for an isolated left latero and basi-cervical mass with no adjacent inflammatory signs, evolving for four years. Imaging (CT and MRI) of the neck revealed a lateral basi-cervical left intermuscular solido-cystic mass without signs of regional infiltration, with permeable cervical vascular axes of normal caliber. A noncontributory cytological examination led to total extra capsular surgical excision of the mass by cervicotomy. It was a retro-jugulo-carotid tumor developed from the right cervical vagus nerve. Histopathological analysis was in favor of a schwannoma.
Discussion: Schwannoma of the vagus nerve is a rare benign tumor, which should be considered in the presence of any isolated laterocervical mass. Preoperative medical imaging (CT and MRI) of the neck represents the essential tool of choice to evoke the diagnosis. The treatment is surgical, in order to confirm the histological diagnosis.
Conclusion: Cervical schwanoma of the vagus nerve is rare, complete extracapsular surgical excision is possible and is the only guarantee of healing.
Keywords: *Schwannoma, vagus nerve, surgery.*

Date of Submission: 26-02-2023 Date of Acceptance: 10-03-2023

I. Introduction

The term 'schwannoma', first introduced in 1935 by Stout, identifies a benign tumor with sporadic malignant degeneration arising from cranial, peripheral and autonomic nerve sheath cells (1). It represents 5% of all benign soft tissue tumors (1), they are the most common type of peripheral nerve tumors (2,3).

Extracranial schwannomas are most commonly found in the parapharyngeal space and involve either the cervical sympathetic trunk or the vagus nerve (4).

Their Diagnosis is still challenging cause of the numerous pathologies the schwannoma can be confused with. Of all the cranial nerves, the vagus nerve has the largest distribution, innervating the neck, thorax, and abdomen, with significant input to the cardiac, respiratory, and digestive systems (5); Schwannomas arising from the vagus nerve present several unique clinical and therapeutic challenges. Injury to the nerve or its peripheral branches during surgical procedures can result in significant patient morbidity affecting voice, swallowing, and respiration.

II. Case Report

We report a case of a 26 year old female patient presented with an isolated left basi-lateral cervical mass without inflammatory signs, evolving for a period of four years. She was asymptomatic, and on examination her cranial nerves were found to be intact and her voice was normal. Computed tomography (CT) and Magnetic resonance imaging (MRI) of the neck demonstrated a latero and basi-cervical left intermuscular solido-cystic mass without signs of loco-regional infiltration, with permeable cervical vascular axes of normal caliber.

A noncontributory cytological examination led to total extra capsular surgical excision of the mass by cervicotomy, It was a retro jugulo-carotid tumor developed at the expense of the right cervical vagus nerve, Several nerves coursing along the capsule were dissected and preserved, Histopathologic examination revealed a schwannoma. The patient experienced no postoperative complications.

III. Discussion

Vagal schwannomas are rare, benign tumors of the head and neck. They usually arise within the third and fifth decades of life, with equal rates among males and females (6). Although surgical resection for large or

symptomatic lesions is the standard of care, nerve sacrifice at the time of resection is associated with significant morbidity.

Due to the rarity of vagal schwannomas, the natural history of these tumors is relatively unclear and can be variable.

The size of the schwannoma could dictate its effects on nerve function, as larger tumors would exert more compression on nerve fibers and potentially cause gradual ischemia (7) Thus, neural deficits may be observed, especially in large tumors that cause local and neural compression (8,9) but still in many cases, the tumor presents as a cervical mass without any symptoms related to nerve's compression similar to our case.

Presenting symptom	Common presenting symptoms of vagal schwannoma
Painless neck mass	Percentage of patients 56.9
Cough Hoarseness Dysphagia Dyspnea	11.6 11.0 6.7 3.7
Vocal cord palsy/paralysis	3.7
Neck pain	3.1
Horner's syndrome	1.8

Table 01: 165 out of 236 cases provided information regarding presenting symptoms (10).
The majority of cases reported a neck mass, including our case.

In addition to the size, the anatomical location of a schwannoma is also important, as smaller tumors may cause neurologic deficits to the affected nerve or surrounding nerves sooner when located in more restricted spaces. The rate of growth for a given schwannoma may also play a role in loss of function. Although most schwannomas have an indolent growth pattern, some tumors exhibit periods of more rapid growth that correlates with symptom progression, contrary to our case, where the mass needed 04 years of growth to reach a significant size.

The most common presentation of vagal schwannoma is a painless, slow-growing neck mass (4,11) , additional symptoms may arise due to the disruption of surrounding anatomy and damage to the vagus nerve(4). (Table 01)

Dysphonia, dysphagia, pharyngeal or airway obstruction, dyspnea, or pain may develop in more advanced cases. 5 additional cranial neuropathies may occur, although this is rare and have only been reported in large tumors (9).

Preoperative diagnosis of schwannoma is difficult because many VNSs do not present with neurological deficits, and several differential diagnosis may be considered, including paraganglioma, malignant lymphoma, branchial cleft cyst, metastatic cervical lymphadenopathy (12) arising from varying locations including carotid body tumors and glomus vagale tumors (4,11).

Imaging studies are crucial in establishing the diagnosis of vagal schwannoma (6), they enable us to first identify the nerve of origin as it has significant implications for preserving neural function during tumor dissection as discussed subsequently.

imaging also enables us to diagnose the type of tumor; CT and MRI suggest the origin of the tumor relating to vascular displacement; usually VNS displaces the internal jugular vein laterally and ICA medially, while schwannoma of the sympathetic chain displaces both together without their separation (13,14).

CT examination of a schwannoma reveals a well-defined fusiform mass with low to intermediate attenuation and variable enhancement. On T2-weighted MR images, schwannomas appear heterogeneously hyperintense, and Iso intense on T1-weighted images 5,12,44 similar to our patient, whom presented with an ovular mass Isointense on T1 weighted images, heterogeneously hyperintense on T2 weighted images, and with intense enhancement after gadolinium injection, the mass also presented a cystic portion.

Ancient schwannomas are differentiated from conventional schwannomas by features resulting from the "aging" of the schwannoma (2,15); Their histology displays degeneration, such as cysts, edema, and fibrosis. They are benign but are sometimes misdiagnosed as malignant due to their heterogeneous contrast enhancement.2 In our patient, the cystic portion can be attributed to the significantly old age of the tumor.

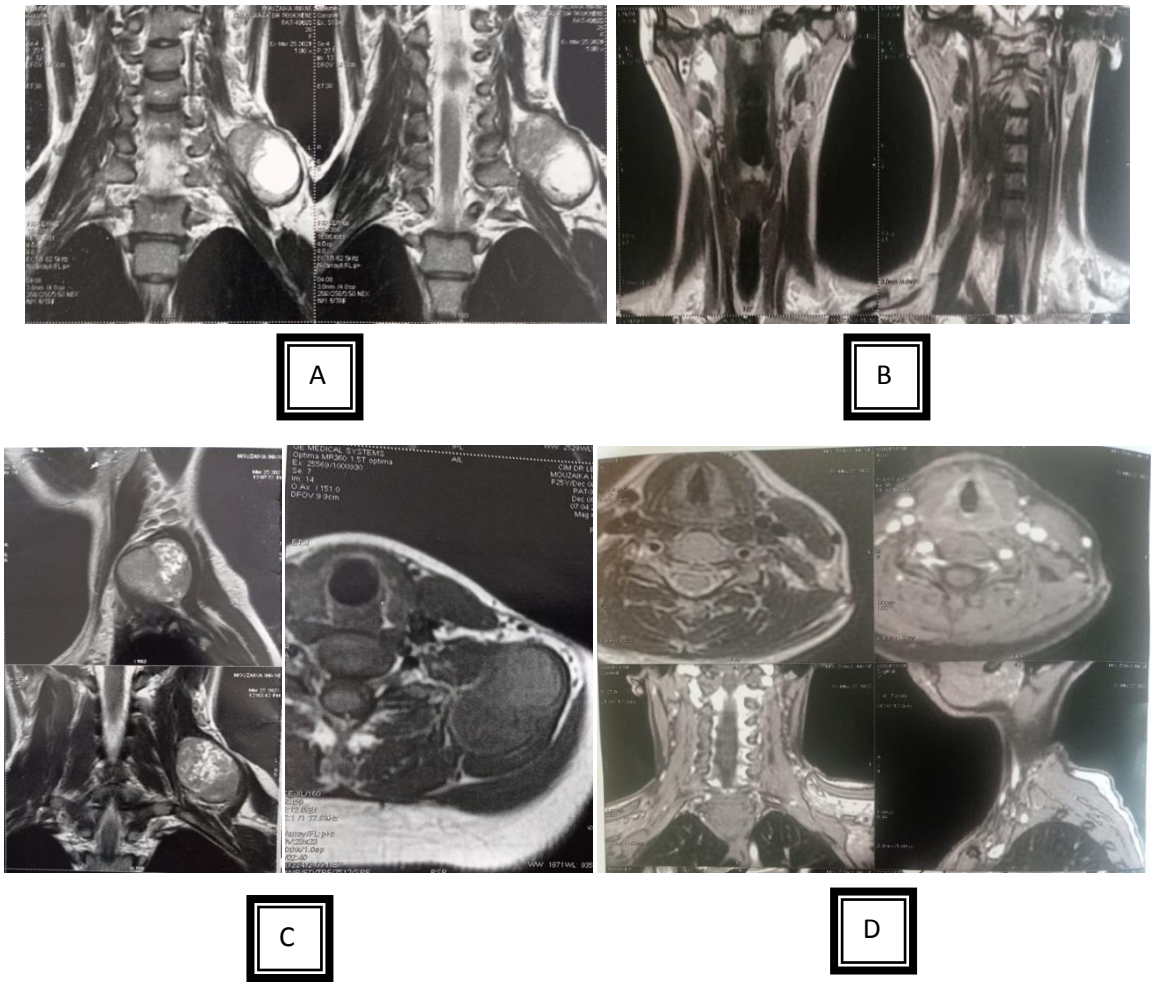

FIGURE 01: Pre (A, C) and Post (B, D) operative MRI images of a vagal schwannoma.
T1-weighted image demonstrates a hypointense neck lesion.
T2-weighted images demonstrate the lesion is heterogeneous to hyperintense.

Many vagal schwannomas are asymptomatic at presentation. Chosing the type of management depends on the size and location of the tumor as well as the age and overall health status of the patient, observation with serial imaging may be a viable option; but surgical excision is the most durable and effective treatment for vagal schwannomas.

All cases in this literature review were eventually treated with surgery, and so did our patient. However, the surgical technique used is somewhat variable. Gibber et al mentioned two different surgical options in patients that have shown tumor growth or developed symptoms (16).

The first is to completely excise the schwannoma with or without nerve sacrifice, depending on whether the tumor can be separated from the nerve. This method removes the schwannoma by cutting the nerve fascicle from which the mass is growing, but often the entire vagus nerve is sacrificed. This method was classified as "gross total resection"

The second option is tumor enucleation, a more conservative, nerve-sparing technique which leaves the tumor capsule behind. In this approach, the nerve fibers are pushed aside, and the capsule of the schwannoma is cut open, allowing for the removal of the schwannoma, leaving the capsule and most involved nerve fibers uncut. This approach was classified as "subtotal resection".

In literature, Intracapsular enucleation was the technique of choice by the majority of surgeons; about one third was extracapsular resections; whereas debulking of tumour was described in just one patient(17). (Table 02) and Almost all of the patients ware approached through a transcervical incision.

In our case we used an extracapsular resection via a transcervical incision, while using nerve stimulation to preserve the vagus nerve functions.

Author	Number of patients	Surgical procedure	Approach	Preoperative symptoms	Vagus resection	Postoperative symptoms	Complete recovery
Hwang [18]	1	Extracapsular resection	Transaxillary	_	_	+	+
Ogawa [19]	1	Extracapsular resection	Transcervical	_	+	+	_
Yasumatsu [20]	10	5Extracapsular resection 5 intracapsular excisions	Transcervical	NR	_	NR	NR
Gibber [21]	1	intracapsular excisions	Transcervical	_	_	_	NR
Bilancia [22]	1	intracapsular excisions	Transcervical	+	_	+	_
Chai [23]	1	intracapsular excisions	Transcervical	+	NR	+	_
Lahoti [24]	4	NR	NR	NR	_	2 + 2 _	NR
Lee [25]	1	Extracapsular resection	Transcervical	_	_	+	_
Liu [26]	6	NR	NR	NR	NR	NR	NR
Sreevatsa [27]	3	intracapsular excisions	NR	_	_	_	_
Kim [28]	6	intracapsular excisions	NR	_	_	1 + 5 _	NR
Chiofalo [29]	1	Extracapsular resection	Transcervical	+	+	+	_
Bahayani [30]	1	Debulking	Transcervical	_	+	+	_
Peyvandi [31]	1	Extracapsular resection	Transcervical	_	_	NR	NR
Biswas [32]	2	NR	NR	NR	_	+	+
Kang [33]	6	intracapsular excisions	Transcervical	NR	_	3_ 3+	+
Lagner [34]	3	NR	NR	NR	NR	NR	NR
Moroni [35]	1	Extracapsular resection	Transcervical	+	_	_	NR
Roh [36]	1	intracapsular excisions	Postauricular	_	_	_	NR
Matsuo [37]	1	NR	Transcervical	+	_	+	_
Ruckert [38]	1	Extracapsular resection	Transcervical	NR	+	NR	_
Giuseppe Cavallaro [17]	2	intracapsular excisions	Transcervical	+	_	1+ 1_	_
Our case	1	Extracapsular resection	Transcervical	_	_	_	+

NR : not reported

Table 02: Surgical features (17)

The first implementation of an IONM system was reported in 1978 by Ojeman et al in the context of vestibular schwannoma surgery (39).

In the case of vagal schwannomas, nerve stimulation during tumor excision can help to distinguish the location of the nerve fibers running over the tumor surface, thereby preserving functional fibers that innervate the larynx and minimizing postoperative morbidities including dysphonia, dysphagia, and a permanent risk of aspiration (40,41).

While operating on our patient and during the tumor dissection, functions of the vagus nerve were continuously monitored; Changes in amplitude were managed with procedure cessation, tissue relaxation, and irrigation until amplitude recovery occurred.

At completion of the surgery, nerve fascicles within the thin schwannoma capsule remained functionally intact to direct stimulation; this technique helped us achieve a satisfactory resection while preserving the absence of symptoms.

Postoperative results vary in a review of the literature and ranged from complete recovery with normal function, similar to our patient; to a variety of deficits that included hoarseness, dysphagia, vocal cord palsy, and vocal cord paralysis. Subtotal resections achieved a higher rate of gross nerve preservation and a higher rate of normal, asymptomatic postoperative course as compared to gross total resection patients (17).

IV. Conclusion

Cervical schwanoma of the vagus nerve is a rare tumor; most commonly presented an asymptomatic cervical mass, complete surgical excision is possible and is the only guarantee of healing. Nerve monitoring techniques are an important tool in the surgical removal of cervical vagal schwannomas, they reduce nerve damage and resulting postoperative complications. Recurrence rates are likely low due to the benign nature of the tumor.

References

[1]. Sinclair CF, Tellez MJ, Tapia OR, Ulkatan S, Deletis V. A novel methodology for assessing laryngeal and vagus nerve integrity in patients under general anesthesia. Clin Neurophysiol 2017;128:1399–1405.
[2]. Colreavy MP, Lacy PD, Hughes J, et al. Head and neck schwannomas—a 10 year review. J Laryngol Otol. 2000;114(02): 119-124. https://doi.org/10.1258/0022215001905058.
[3]. Bhattacharyya AK, Perrin R, Guha A. Peripheral nerve tumors: management strategies and molecular insights. J Neurooncol. 2004; 69(1–3):335-349. https://doi.org/10.1023/B:NEON.0000041891. 39474.cb
[4]. Behuria S, Rout T, Pattanayak S. Diagnosis and management of schwannomas originating from the cervical vagus nerve. Ann R Coll Surg Engl. 2015;97(2):92-97. https://doi.org/10.1308/ 003588414X14055925058355.
[5]. Câmara R, Griessenauer CJ. Anatomy of the vagus nerve. Nerves Nerve Inj. 2015;1:385-397. https://doi.org/10.1016/B978-0-12-410390-0.00028-7
[6]. Chiofalo MG, Longo F, Marone U, Franco R, Petrillo A, Pezzullo L. Cervical vagal schwannoma. A case report. Acta Otorhinolaryngol Ital. 2009;29(1):33-35
[7]. Liu HL, Yu SY, Li GKH, Wei WI. Extracranial head and neck schwannomas: a study of the nerve of origin. Eur Arch Otorhinolaryngol. 2011;268(9):1343-1347. https://doi.org/10.1007/ s00405-011-1491-4.
[8]. De Araujo CEN, Ramos DM, Moyses RA, Durazzo MD, Cernea CR, Ferraz AR. Neck nerve trunks schwannomas: clinical features and postoperative neurologic outcome. Laryngoscope. 2008; 118(9):1579-1582. https://doi.org/10.1097/MLG.0b013e31817b0702.
[9]. Kanatas A, Mücke T, Houghton D, Mitchell DA. Schwannomas of the head and neck. Oncol Rev. 2009;3(2):107-111. https://doi.org/ 10.1007/s12156-009-0015-6
[10]. Mykayla L. Sandler BA1 | John R. Sims MD2 | Catherine Sinclair MD2 | Kayvon F. Sharif BA1 et Al. Vagal schwannomas of the head and neck: A comprehensive review and a novel approach to preserving vocal cord innervations and function DOI: 10.1002/hed.25758
[11]. Furukawa M, Furukawa MK, Katoh K, Tsukuda M. Differentiation between schwannoma of the vagus nerve and schwannoma of the cervical sympathetic chain by imaging diagnosis. Laryngoscope. 1996;106(12 Pt 1):1548-1552. http://www.ncbi.nlm.nih.gov/pubmed/8948621
[12]. Colreavy MP, Lacy PD, Hughes J, et al. Head and neck schwannomas e a 10-year review. J Laryngol Otol 2000;114:119e24.
[13]. Cavallaro G, Pattaro G, Lorio O, et al. A literature review on surgery for cervical vagal schwannomas. World J Surg Oncol 2015;13:130
[14]. Furukawa M, Furukawa MK, Katoh K, et al. Differentiation between schwannoma of the vagus nerve and schwannoma of the cervical sympathetic chain by imaging diagnosis. Laryngoscope 1996;106(12 pt.1):1548e52.
[15]. Shilpa B. Case report ancient schwannoma—a rare case. Ethiop J Health Sci. 2012;22(2):215-218
[16]. Gibber MJ, Zevallos JP, Urken ML. Enucleation of vagal nerve schwannoma using intraoperative nerve monitoring. Laryngoscope. 2012;122(4):790-792. https://doi.org/10.1002/lary.22485
[17]. Giuseppe Cavallaro, Giada Pattaro, Olga Iorio, Marcello Avallone and Gianfranco Silecchia A literature review on surgery for cervic schwannomas, Cavallaro et al. World Journal of Surgical Oncology (2015) 13:130 DOI 10.1186/s12957-015-0541-6
[18]. Hwang KR, Kim JW, Kim HK, Lee SW. A cervical vagal schwannoma mimicking a parathyroid cyst. Clin Exp Otorhinolaryngol. 2014;7:153–6
[19]. Ogawa T, Kato T, Ikeda A, Nishimura K, Tsuchiya Y, Okamoto H, et al. Case of malignant transformation of vagus nerve schwannoma to angiosarcoma. Head Neck. 2014;36:E17–20.
[20]. Yasumatsu R, Nakashima T, Miyazaki R, Segawa Y, Komune S. Diagnosis and management of extracranial head and neck schwannomas: a review of 27 cases. Int J Otolaryngol. 2013;2013:973045.
[21]. Gibber MJ, Zevallos JP, Urken ML. Enucleation of vagal nerve schwannoma using intraoperative nerve monitoring. Laryngoscope. 2012;122:790–2
[22]. Bilancia R, Ampollini L, Cattelani L, Carbognani P, Rusca M. Schwannoma of the cervical vagus nerve. Ann Thorac Surg. 2011;91:e13
[23]. Chiun KC, Tang IP, Prepageran N, Jayalakshmi P. An extensive cervical vagal nerve schwannoma: a case report. Med J Malaysia. 2012;67:342–4
[24]. Lahoti BK, Kaushal M, Garge S, Aggarwal G. Extra vestibular schwannoma: a two year experience. Indian J Otolaryngol Head Neck Surg. 2011;63:305–9
[25]. Lee RM, Ong CP, Jacobsen AS, Chan MY, Hwang WS. Malignant peripheral nerve sheath tumor mimicking carotid body tumor–case report and review. J Pediatr Surg. 2011;46:554–8
[26]. Liu H, Yu S, Li GK, Wei WI. Extracranial head and neck schwannomas: a study of the nerve of origin. Eur Arch Otorhinolaryngol. 2011;268:1343–7
[27]. Sreevatsa MR, Srinivasarao RV. Three cases of vagal nerve schwannoma and review of literature. Indian J Otolaryngol Head Neck Surg. 2011;63:310–2
[28]. Kim SH, Kim NH, Kim KR, Lee JH, Choi HS. Schwannoma in head and neck: preoperative imaging study and intracapsular enucleation for functional nerve preservation. Yonsei Med J. 2010;51:938–42
[29]. Chiofalo MG, Longo F, Marone U, Franco R, Petrillo A, Pezzullo L. Cervical vagal schwannoma. A case report. Acta Otorhinolaryngol Ital. 2009;29:33–5
[30]. Bhayani MK, MacCracken E, Frim D, Baroody FM. Prolonged cricopharyngeal muscle spasm after resection of the cervical vagus nerve in a 15-year-old. Pediatr Neurosurg. 2008;44:71–4.
[31]. Peyvandi A, Samadian M, Ahmady-Roozbahany N. Photoclinic. What is your diagnosis? Photoclinic diagnosis: vagus nerve schwannoma. Arch Iranian Med. 2008;11:669–71

[32]. Biswas D, Marnane CN, Mal R, Baldwin D. Extracranial head and neck schwannomas - a 10-year review. Auris Nasus Larynx. 2007;34:353–9
[33]. Kang GC, Soo KC, Lim DT. Extracranial non-vestibular head and neck schwannomas: a ten-year experience. Ann Acad Med Singapore. 2007;36:233–28.
[34]. Langner E, Del Negro A, Akashi HK, Akashi HK, Costa Araùjo PP, Tincani AJ, et al. Schwannomas in the head and neck: retrospectives analysis of 21 patients and review of the literature. Sao Paulo Med J. 2007;125:220–2.
[35]. Moroni AL, Righini C, Faure C, Serra-Tosio G, Lefournier V. Features of an unusual calcified schwannoma of the superior laryngeal nerve. AJNR Am J Neuroradiol. 2007;28:981–2
[36]. Roh JL. Resection of cervical vagal schwannoma via a post-auricular approach. Acta Otolaryngol. 2006;126:318–20.
[37]. Matsuo R, Kamouchi M, Inoue T, Okada Y, Ibayashi S. Cerebral infarction due to carotid occlusion caused by cervical vagal neurilemmoma: report of a case. Stroke. 2002;33:1428–31
[38]. Rückert RI, Fleige B, Rogalla P, Woodruff JM. Schwannoma with angiosarcoma. Report of a case and comparison with other types of nerve tumors with angiosarcoma. Cancer. 2000;89:1577–85.
[39]. Ojemann RG, Levine RA, Montgomery WM, McGaffigan P. Use of intraoperative auditory evoked potentials to preserve hearing in unilateral acoustic neuroma removal. J Neurosurg. 1984;61(5): 938-948. https://doi.org/10.3171/jns.1984.61.5.0938.
[40]. Netterville JL, Jackson CG, Miller FR, Wanamaker JR, Glasscock ME. Vagal paraganglioma. Arch Otolaryngol Head Neck Surg. 1998;124:1133-1140
[41]. Ijichi K, Kawakita D, Maseki S, Beppu S, Takano G, Murakami S. Functional nerve preservation in extracranial head and neck schwannoma surgery. JAMA Otolaryngol Head Neck Surg. 2016;142(5):479-483. https://doi.org/10.1001/jamaoto. 2016.0113

Kamel Bouaita, et. al. " Cervical cystic schwannoma of the vagus nerve Case report and review of literature." *IOSR Journal of Dental and Medical Sciences (IOSR-JDMS)*, 22(3), 2023, pp.36-41.