CASE REPORT

Cervical Vagal Schwannoma: A Case Report

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Nerve sheath tumors arising from the cervical vagus nerve are rare. These tumors mostly present as asymptomatic, slowly growing masses in the lateral neck. The authors reported a case of vagal schwannoma in a healthy male patient with a small lump in his neck. The neck examination revealed a firm, non-tender, non-pulsatile mass measuring 3×3 cm in the upper part of the right neck at the level of the carotid bifurcation. No neurologic deficit was detected. Fine needle aspiration showed inconclusive results. The MRI findings were distinctive, enabling a diagnosis before surgery. The preferred treatment option was gross total resection while preserving the vagus nerve. Although the authors meticulously dissected the tumor from the nerve and the nerve trunk was preserved, postoperative complications of this case were ipsilateral vocal cord paralysis and hypoglossal nerve palsy. Complete surgical removal is crucial to prevent recurrence. The prognosis of the complete cure for benign neurogenic tumors in the neck is excellent.

Keywords: Nerve sheath tumor; Schwannoma; Vagus nerve

Received 25 June 2024 | Revised 24 September 2024 | Accepted 9 October 2024

J Med Assoc Thai 2024;107(11):920-5

Website: http://www.jmatonline.com

Cervical vagal schwannoma is a rare, slow-growing tumor usually reported in individuals aged 30 to 50 years, though it can occur at any age[1]. There is no gender-related predisposition[1,2]. It is usually asymptomatic benign lesion[1,2]. Imaging tests assist in diagnosing vagal nerve neoplasm, with magnetic resonance imaging (MRI) being the most sensitive and specific study available. MRI provides important preoperative information useful in planning surgical treatment[2]. Complete surgical resection is the treatment of choice[3]. The present case report was approved by the Ramathibodi Hospital Institution Review Board (MURA2024/453), and written informed consent was obtained.

Case Report

A 28-year-old Thai male presented with a gradually increasing right upper neck mass for three months. He had been in good health prior to this condition. He initially visited a private hospital with a small lump in his neck. He denied experiencing any pain, hoarseness, dysphagia, aural fullness, weight loss, dyspnea, epistaxis, blurred vision, and aspiration. No signs of neurological deficit were detected. There was no paralysis of any cranial nerves on the involved side. Horner syndrome was absent. He denied any tobacco or alcohol use. There were no superficial skin changes on his neck. Examination revealed a firm, non-tender, and non-pulsatile mass measuring 3×3 cm in the upper right neck at the level of carotid bifurcation (Figure 1). The carotid pulse was palpable, and no carotid bruit was observed. The

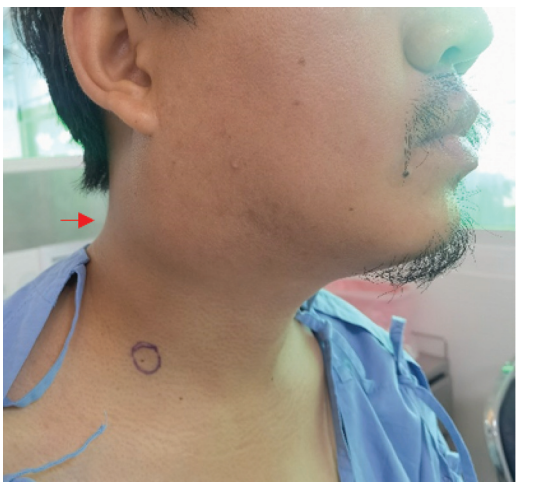

Figure 1. The neck examination revealed a palpable mass at level II on the right side of the neck. The arrow indicates the position of the mass.

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How to cite this article:

Leelapatharaphan N, Temtrirath K. Cervical Vagal Schwannoma: A Case Report. J Med Assoc Thai 2024;107:920-5.

DOI: 10.35755/jmedassocthai.2024.11.920-925-01184

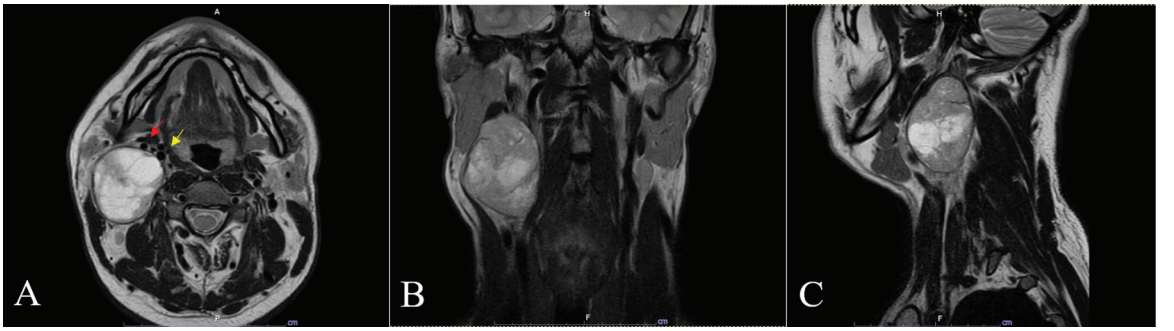

Figure 2. MRI pic revealed well-circumscribed mixed solid cystic lesion seen iso to hyperintense T2 mass with heterogeneous enhancement and cystic component at the right cervical level II, measuring about 3.5×3.1×4.7 cm in AP×TRV×CC dimensions. (A) Axial view: The mass displaced proximal right ICA/ECA anteriorly. The red arrow indicates the ECA, while the yellow arrow indicates the ICA. (B) Coronal view. (C) Sagittal view.

mass was slightly movable.

His previous doctor sent him for ultrasound of the neck, which showed a 3.2×2.7×4.9 cm soft tissue mass with central necrosis abutting aortic bifurcation. A fine needle aspiration (FNA) test revealed a spindle cell tumor favoring pleomorphic adenoma. Tissue for histopathology was recommended. AFB was negative. A contrast-enhanced computed tomography (CT) of the neck revealed an oval-shaped, sharply circumscribed, heterogeneous enhancing mass epicenter at right carotid space with thick irregular peripheral enhancement and central low attenuation (measured about 4.3×2.8×3.5 cm). Differential diagnoses were nerve sheath tumor and lymph node enlargement. There were no other abnormalities in the neck.

He was referred to the authors' hospital for proper management. He arrived at the Otolaryngology outpatient clinic at Ramathibodi Hospital with his CT results. Indirect laryngoscopy was performed to confirm normal vocal cord mobility on both sides. Repeated ultrasound-guided fine needle aspiration showed neoplasm with basaloid feature (Milan IV-B: salivary gland neoplasm with uncertain malignant potential). After the aspiration, he did not experience any symptoms such as hoarseness or hematoma. The neck MRI result revealed no significant change in overall size of the pre-existing well-circumscribed mixed solid cystic lesion seen as hypo to isointense T1 iso to hyperintense T2 mass with heterogeneous enhancement and cystic component at the right cervical level II, measuring about 3.5×3.1×4.7 cm, respectively. The lesion lied deep to right sternocleidomastoid muscle (SCM), displaced the distal right common carotid artery and proximal right internal carotid artery (ICA) or external carotid artery (ECA) anteriorly and compressed the right internal jugular vein (IJV) and separated it from ICA and ECA. No definite synchronous lesion along the other areas of the visualized neck was seen. MRI suggested an enhanced mass with a cystic component at right carotid space, most likely vagal schwannoma (Figure 2).

The authors planned to do a total resection of the mass, but the operation was postponed due to COVID-19 situation. While waiting two years for the operation, the mass had progressively increased in size. The physical examination revealed a firm mass measuring 6×8 cm. There was no tongue deviation, and the tonsils were normal in size with no lateral pharyngeal wall bulging. He had no cough while palpating the mass. He had normal eye examination with no ptosis and pupil reacted normally to light.

The aim of the treatment was gross total resection with preservation of the vagus nerve. Consent for this operation was obtained, including details of specific complications such as temporary or permanent vagus nerve injury, hypoglossal nerve injury, and Horner syndrome. He underwent exploration and resection of the tumor. The patient had a complete surgical excision of the mass under general anesthesia. The authors did not use an intraoperative nerve monitoring device. The incision was done horizontally, 2 cm below the mandible, through the platysma muscle. The subplatysmal flap was elevated. The anterior end reached the anterior border of the sternocleidomastoid muscle. The posterior end was extended to the posterior border of the tumor. The tumor was located deep within the sternocleidomastoid muscle, at its anterior border (Figure 3). The feeding artery to the tumor was clamped and cut. The tumor was found in the carotid sheath. The IJV was identified lateral to the tumor and dissected from the tumor. The ICA and ECA were medial to the tumor. The tumor separated

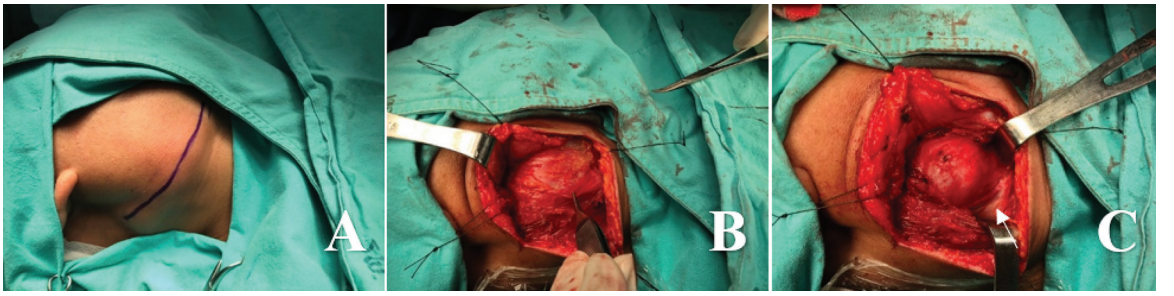

Figure 3. (A) Incision was 2 cm below mandible. (B) The tumor was located deep within the SCM, and the forceps picked up the anterior border of the SCM. (C) The white arrow indicates IJV.

between ICA, ECA anteriorly, and IJV posteriorly.

Even though an extracapsular technique was performed under loupe assistance, the tumor was accidentally ruptured, releasing 1 mL of clear brown content. The tumor was well encapsulated and not infiltrative in character. The tumor was meticulously dissected from the nerve. Mobilization of the tumor revealed that its sheath was adhered anteriorly to the vagus nerve. The nerve trunk was preserved (Figure 4). The tumor was freed from the surrounding tissue by sharp and blunt dissection. The resection of the tumor was achieved with the vagus nerve in continuity.

The mass was red to brown in color, firm consistency, well encapsulated and featured a smooth surface (Figure 5).

The drain was inserted. The subcutaneous tissue was closed with absorbable surgical sutures and the wound was closed in layers with non-absorbable surgical nylon sutures. The pathological results from the permanent section indicated characteristics of schwannoma, which consisted of Antoni A and Antoni B areas (Figure 6).

Immediately postoperative, the patient developed dysphonia after being awake from anesthesia. The bedside examination of the larynx with a fiberoptic laryngoscope confirmed paralysis of the right vocal cord and normal movement of the left vocal cord. He had a slight tongue deviation to the right side. He denied any choking, coughing, or aspiration. The ophthalmologist detected no ptosis or miosis of the eye. At the six-month follow-up, the patient was well except for experiencing hoarseness and a minimal tongue deviation to the right side. He underwent a fat graft medialized laryngoplasty with a satisfactory outcome. His voice had improved significantly, allowing him to communicate with greater confidence. At the two-year follow-up, examinations revealed no recurrence or new lesions.

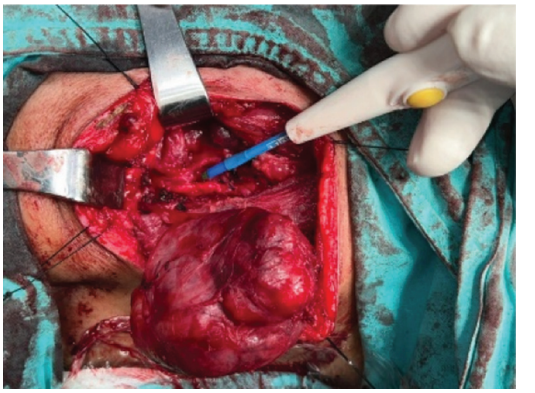

Figure 4. Mobilization of the tumor revealed that its sheath was adhered anteriorly to the vagus nerve. The nerve was preserved and identified at the tip of the cautery device.

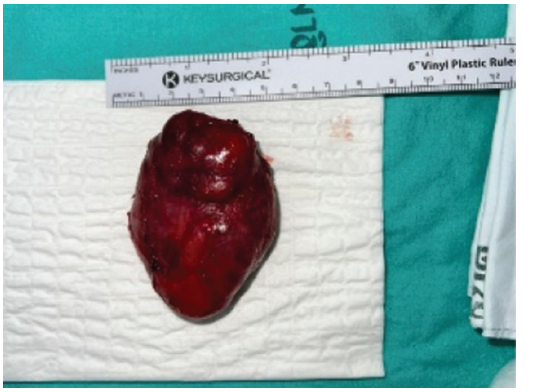

Figure 5. The gross appearance of the tumor.

His voice recovered to near normal, but his tongue still slightly deviated to the right.

Discussion

Schwann cells form the sheath around the nerve fibers. A benign tumor arising from the cells of Schwann of the neurilemma covering the nerve is called the neurilemoma or schwannoma[3]. It is

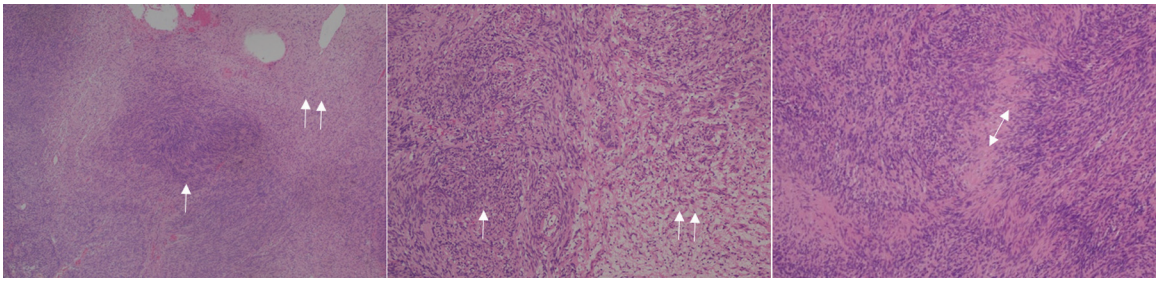

Figure 6. Histological images displaying characteristic features of schwannoma, including Antoni A and Antoni B areas (40X magnification).

→ Antoni A consist of palisading of the nuclei around a central mass of cytoplasm called Verocay bodies.
⇉ Antoni B contain a loose stroma with no distinct pattern by the fibers and cells.
↔ Verocay bodies are the central mass of cytoplasm surrounded by nuclei that are palisading in Antoni A areas.

the most common type of benign peripheral nerve tumor in adults. In 1908, Verocay had first described histologically the microscopic appearance of this specific well-differentiated tumor, which developed within the nerve sheath. In 1935, Stout proposed the term "neurilemoma" for benign nerve sheath tumors. During that time, the cell of origin was uncertain[4]. In 1940, Murray and Stout cultivated the tumor in vitro and demonstrated by tissue cultures that the cells of these nerve sheath tumors were morphologically and physiologically similar to the cells of Schwann[5]. After their experimental work, it was concluded and agreed to use "schwannoma" to denote solitary peripheral nerve sheath tumors with a benign course[6]. Most of the schwannomas, or 75% of the cases, had occurred in the extremities, mediastinum, chest, and abdominal cavities[3]. Approximately 25% of the schwannomas reported in the head and neck. From the case series, there were mainly the vagus nerve, followed by the cervical sympathetic nerve, and lastly, the brachial plexus[7]. Among them, 11 (58%) were from the vagus nerve, 7 (37%) from the cervical sympathetic chain, and one (4%) from the brachial plexus C5 nerve root[7]. Vagal schwannoma typically presents between the third and fifth decades of life but may occur at any age[1] and is equally prevalent among both sexes. In the case series, schwannomas presented in a broad age group. The age range was from 22 to 74 years presenting with this disease[7]. However, a study by Shrikrishna et al. found more prevalence in males, at a ratio of 3:1, with the highest incidence in the second decade of life[8]. No causative factors are known. The most frequent complaints of vagus nerve schwannoma are neck mass or localized swelling in the neck[9]. Vagal schwannoma typically appears as a firm, ovoid, or fusiform mass in the lateral neck, with a history of gradual enlargement over several months to years. The tumor usually causes no symptoms and pain is absent[1,3]. Occasionally, symptoms referring to vagal irritation are presented and manifested by coughing, hoarseness, or mild tachycardia[9]. If a mass is large and located superiorly in the neck, it may present as a bulge in the lateral pharyngeal wall within the parapharyngeal space. The tumor can sometimes be mistaken for a carotid body tumor, which usually presents with pulsation or bruit and is closely attached to the carotid artery[1].

Typically, schwannomas occur unilaterally. However, in the case series, two patients had neurofibromatosis and presented with schwannomas in more than one site, one of them was located in head and neck[7]. In around 5% of the cases, they were multiple and seen with neurofibroma. Rarely, benign schwannomas may undergo malignant transformation. Stout, in 1935, presented a case of malignant paraganglioma of the ganglion nodosum of the vagus nerve[4]. Therefore, malignant tumors of the vagus nerve are possible. It is conceivable that malignant tumors of the vagus nerve would produce more symptoms of vagal irritation with invasion and destruction of the nerve fibers[9]. Vagal schwannoma is uncommon, and the differential diagnosis is challenging. Some of the lesions that would necessarily be considered in the differential diagnoses are lipoma, submandibular gland tumor, reactive or metastatic cervical lymphadenopathy, lymphoma, carotid body tumor, carotid artery aneurysm, cystic hygroma, thyroid adenoma, branchial cleft cyst, sebaceous cyst, tuberculous lymph node, parotid tumor, and paraganglioma[9].

Lipoma and branchial cleft cyst are usually softer than schwannoma. Thyroid adenoma is lower in the neck and generally not under the SCM. Tuberculous

lymph node can present with a lump at the neck but may occur with dimpling of the neck and drainage tracts[1].

Salivary gland tumor, cervical lymphadenopathy, and lymphoma must be included in the differential diagnoses, as these conditions often cannot be differentiated without histopathological examination. FNA is often inconclusive but imaging tests are helpful. In a case series of 25 patients with schwannoma in the head and neck region, FNA was reported as consistent with a benign nerve sheath tumor in 50% of the cases and was inconclusive in the rest of the cases due to acellular smear[7]. A retrospective study by Bondi et al., which included 18 patients and half of the group, underwent ultrasonography with FNA, which was the diagnostic tool in 30% of the cases[2]. A CT scan is usually adequate for an appropriate diagnosis, but an MRI is the gold standard for investigation. From the previous study, MRI was the diagnostic tool in 77% of the cases[2]. Imaging studies, particularly MRI, will suggest the diagnosis, revealing low signal intensity on T1-weighted images and high signal intensity on T2-weighted images[10]. A retrospective review of radiographic cross-sectional images of parapharyngeal schwannomas found that schwannomas arising from cervical sympathetic chains displaced both carotid and jugular vessels without separating them. In contrast, carotid and internal jugular vessels were separated in the case of vagal schwannomas. This also aided in distinguishing vagal schwannomas from those originating in the cervical sympathetic chain[11]. Schwannoma of vagus origin displaces the IJV laterally and carotid artery medially[12]. If MRI shows ''salt and pepper'' appearance, it suggests that that it is more likely to be paraganglioma. Schwannoma can be distinguished from aneurysm by noting that an aneurysm exhibits a vascular flow void on imaging. Malignant conditions show lytic lesions and invasion, which are not seen in schwannoma[13].

Complete surgical excision of the tumor is the treatment of choice. In the surgery, every effort should be made to preserve the continuity of the nerve pathway of the involved nerve and the associated nerves in that area[3]. The primary goal of treatment is complete tumor removal with preservation of function.

Vagal schwannoma is situated beneath the upper one-third of the sternocleidomastoid muscle[9]. It is not attached to surrounding structures. Careful dissection of the tumor from its nerve of origin, while taking care to preserve neurological pathways, should be performed. Intraoperative nerve monitoring can be applied to vagus nerve identification to reduce the risk of injury. Case reports using this technique showed promising results but need more prospective studies[14,15]. The critical anatomic point is that schwannoma arises from a solitary entering nerve fascicle at the proximal pole. Therefore, gross total removal can be accomplished by sacrificing the entering and exiting fascicles, causing functional loss. End-to-end anastomosis or nerve grafting may be mandated in exceptional cases where nerve preservation is not possible[16].

Vocal cord paralysis is the most common postoperative complication. If the surgeon had to excise the vagus nerve at a high level in the neck, it would result in dysphagia, paralysis of the ipsilateral vocal cord, and soft palate. Ipsilateral anesthesia of the pharynx and larynx was also found[3]. In a case series from Doseman et al., 23 cases in that study had no significant intraoperative or immediate postoperative complications. However, one patient with vagal schwannoma developed ipsilateral vocal cord palsy along with hypoglossal nerve palsy in the postoperative period. Another patient with vagal schwannoma developed ipsilateral vocal cord palsy with hypoglossal nerve palsy as well as right marginal mandibular nerve weakness[7].

Histologically, two distinct types of cellularity were identified, a classification first described by Antoni in 1920[10]. Characteristic Antoni A or B cells in histopathology confirmed the diagnosis in all the cases. Immunohistochemistry (IHC) using S-100 protein had been shown to be beneficial in confirming diagnosis in uncertain cases[7]. Complete surgical excision of the tumor is essential to prevent recurrence[9]. The prognosis of the complete cure for benign neurogenic tumors in the neck is excellent.

Conclusion

Complete surgical excision of vagal schwannoma is the treatment of choice. Preventing postoperative complications, such as vagus nerve injury, remains a significant challenge in this operation.

What is already known on this topic?

Recommended treatment of vagal schwannoma is usually surgical excision but because of the intimate relationship to the nerve of origin, it is often difficult to preserve the function of the nerve.

What does this study add?

After the tumor was excised with nerve

preservation, the patient had dysphonia and deviated tongue. Therefore, patient counseling is important regarding the potential risks and morbidities of surgical intervention. Careful microsurgical dissection combined with intraoperative nerve monitoring would lead to better outcomes in terms of postoperative morbidity.

Conflicts of interest

The authors declare no conflict of interest.

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