

Rare Presentation of Cervical Vagal Schwannoma in Adolescence: A Case Report

Demah Alayadhi¹, Abdullah Arafat^{2*}, Mohammed Elkarim², Mohammed Alessa², Ali Alassiri³

¹ College of Medicine, King Saud Ben Abdulaziz University of Health Sciences.

² Department of ORL-H&N Surgery, King Abdulaziz Medical City, National Guard Health Affairs.

³ Department of Pathology and Lab Medicine, King Abdulaziz Medical City, National Guard Health Affairs.

Received: December 06, 2017; Accepted: December 13, 2017; Published: December 21, 2017

***Corresponding author:** Abdullah Arafat, Department of ORL-H&N Surgery, King Abdulaziz Medical City, National Guard Health Affairs; Saudi Arabia. E-mail-
Arafat.As@outlook.sa

Abstract

Schwannoma is a rare benign tumor arising from the Schwann cells that produce myelin sheath that cover cranial, peripheral and autonomic nerves. Cervical vagal schwannomas are slow growing asymptomatic solitary benign neck tumors. It affects females and males equally, and it is most common between third and fifth decades of age. Cervical vagal schwannoma requires high clinical suspicion to be diagnosed because of its rarity. Imaging and FNA are a valuable investigation to diagnose and differentiate cervical vagal schwannoma from other diseases. The treatment of choice is complete surgical resection of the mass and histopathology of the excised mass is obligatory for a definitive diagnosis. We report a case of a 17-year-old male patient presented with left neck mass who was diagnosed with cervical schwannoma of vagal nerve origin, which is rare.

Introduction

Schwannomas, also known as neuromas, neurinomas, or neurilemmomas a rare benign tumor arising from the Schwann cells that produce myelin sheath which covers nerve fibers [1,2]. It can originate from any cranial, peripheral and autonomic nerves of the body except olfactory and optic nerves [13]. It has been stated that schwannoma represents 5% of all soft tissue tumors [4]. About one third of schwannoma is diagnosed in the head and neck region [3]. Of the reported cases, approximately half of the parapharyngeal schwannomas arise from the vagus nerve [8]. Cervical vagal schwannomas are slow growing benign tumors. Almost always, manifest as asymptomatic solitary lateral neck mass that can be palpated along the medial border of sternocleidomastoid muscle [8]. It affects females and males equally, and it is most common between third and fifth decades [5]. Cervical vagal schwannoma requires high clinical suspicion to be diagnosed because of its rarity [8]. In addition, imaging is important to diagnose and differentiate vagal schwannoma from other disease [8]. The treatment of choice is complete surgical resection of the mass and histopathology of the excised mass is obligatory for a definitive diagnosis [5].

Case Report

A 17-yearsold male presented to our center with progressive, slow growing left neck mass which was noticed by his family 4 years ago. He had no obstructive symptoms, dyspnea, dysphagia, or other associated symptoms nor was there a history of radiation or family history of neck tumors. Examination revealed hard, pulsatile, nontender mass placed in the carotid space measured 3 x 4 cm with normal overlying skin and no discharge. It was movable in horizontal but not in vertical plane. There were no palpable lymph nodes and the facial nerve was intact. Examination of the ear, nose and throat was unremarkable. Flexible nasopharyngolaryngoscopy showed normal bilateral vocal cord mobility. Ultrasound guided biopsy of the mass showed benign spindle cell tumor consistent with schwannoma, which was confirmed by immunochemistry and was negative for malignancy. On MRI, there was a well-defined homogenous enhanced mass displacing common carotid artery and internal jugular vein posteriorly and laterally demonstrating intense signal on T1 WI, Hyper intense signal on T2 WI [Figures 1,2]. Subsequent contrasted CT revealed a well-defined oval mass involving the left carotids sheath showing heterogeneous faint enhancement with mild displacement of adjacent vascular structures [Figure 3].

Based on patient's presentation, examination and investigations, the patient was operated by transverse left cervical skin incision. Dissection of the all muscles was done. A yellowish mass measuring 3 x 4 cm was seen in the area between the carotid artery and internal jugular vein. Tumor was excised completely from the left vagus nerve, leaving the nerve intact. The diagnosis of schwannoma was confirmed by histopathology which reveals a schwannoma composed of fascicles of plump spindle cells with elongated tapering nuclei and moderately abundant, eosinophilic cytoplasm with indistinct cell borders. The cellularity is variable due to the characteristic zonation of alternating hypercellular areas (Antoni A) figure 4a, with distinctive focal nuclear palisading surrounding aggregates of cellular processes (Verocaybodies)

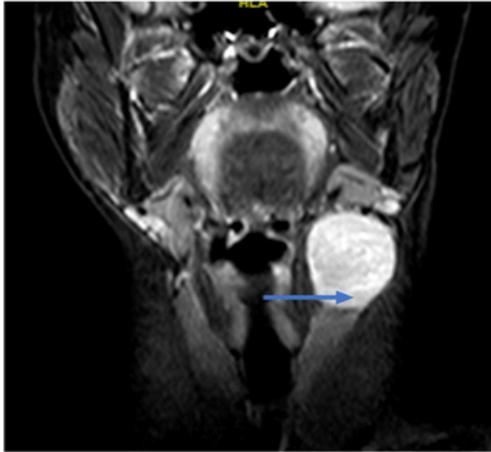


Figure 1: MRI of the neck, coronal section, T2WI: Well-circumscribed oval-shaped Hyperintense mass.

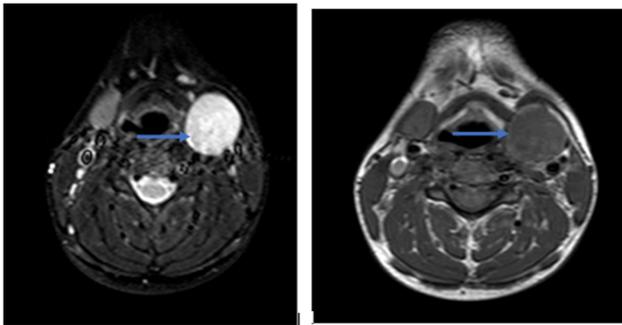


Figure 2: MRI of the neck, axial section: Well-circumscribed oval-shaped mass, Hyperintensity in T2WI (on the top) and hypointensity T1WI (on the bottom).



Figure 3: Contrast Computed Tomography of the neck, coronal section: Well-circumscribed oval-shaped which displace the CA and IJV posteriorly and laterally

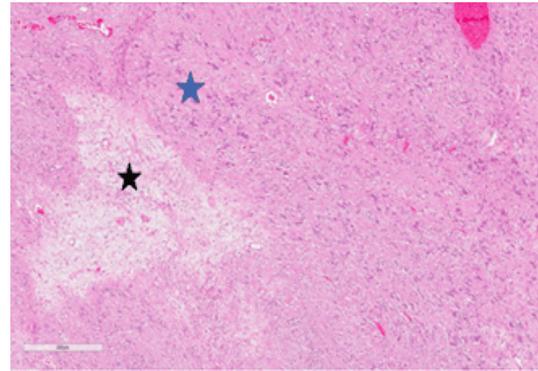


Figure 4A: Low power view showing alternating areas of Antoni A (hypercellular; blue star) and Antoni B (hypocellular; black star)

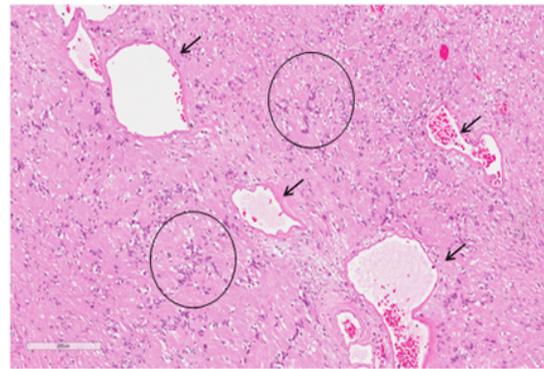


Figure 4B: Verocay body (circles), a distinctive feature of schwannoma resulting from regimentation of Schwann cell nuclei with nuclear free zone on either side, also seen hyalinized vasculature which is a common feature (arrows).

figure 4b, and looser hypocellular areas (Antoni B) figure 4b [Figure 4a, Figure 4b]. Thick-walled, hyalinized blood vessels are also present figure 4b. Patient was followed as an outpatient at 2 weeks, 6 weeks, 3 months, 4 months, 8 months and 1 years. Patient developed mild left ptosis 2 weeks postoperatively, which was recovered spontaneously after 3 months and both vocal cords were intact in post-operative follow up.

Discussion

Schwannoma is a benign tumor arising from the Schwann cells that produce myelin sheath that cover cranial, peripheral and autonomic nerves [1,2]. It presents as encapsulated, well circumscribed, firm, nodular lesions [2]. It has been stated that schwannoma represent 5% of all soft tissue tumors [4]. About one third of schwannoma is diagnosed in the head and neck region, and the most common involves cranial nerve is the eighth nerve, called vestibular schwannoma (3). Compared to vestibular schwannoma, cervical vagal schwannoma is a rare tumor [5]. Cervical vagal schwannomas are slow growing tumors that adhere to their nerve of origin [6]. Majority of vagal nerve schwannomas are benign, and malignant transformation is rare [6]. Most of patients with cervical vagal schwannoma

manifest as asymptomatic solitary lateral neck mass [8]. The most common presenting symptom is hoarseness, but the most specific sign for cervical vagal schwannoma is paroxysmal cough [8]. Paroxysmal cough is elicited by stimulation of the vagal nerve during mass palpation [4]. Carotid Artery (CA) is displaced anteriorly and medially by cervical vagal schwannomas while Internal Jugular Vein (IJV) is displaced posteriorly and laterally [6]. The mass is almost always mobile in the horizontal but not in vertical direction [6]. The differential diagnoses of the mass could be paraganglioma, branchial cleft cyst, carotid body tumor, malignant lymphoma, metastatic cervical lymphadenopathy, and cervical vagal schwannoma [9]. Preoperative diagnosis of cervical vagal schwannoma is difficult because of its rarity [3]. FNA and imaging can be helpful preoperatively to diagnose and differentiate cervical vagal schwannoma from the other diseases [3]. FNA value is questionable since the treatment of choice is a complete surgical resection of the mass [9]. MRI is considered the gold standard for cervical vagal schwannoma, whereas CT findings usually nonspecific and not helpful [1,9]. On MRI, the lesion is heterogeneously hyperintense in T2WI and hypointense in T1WI [10]. After gadolinium administration, the lesion showed heterogeneous hyperintensity [10]. MRI findings are important to differentiate schwannomas by its origin in relation to vascular displacement. The vagus nerve schwannomas displace CA medially and IJV laterally, where schwannoma of the sympathetic chain leads to displacement of these vessels without separation [5,1]. Also, MRI is useful to differentiate tumors of carotid body which splay the carotid bifurcation; lyre sign [1]. In addition, MRI is used to exclude paragangliomas which have salt and pepper sign, from schwannomas. So, salt represents the hyperintense (white part) of mass due to hemorrhage in the hyper vascular tumors, where pepper represents hypointense (black part) of the mass due to increase blood flow in the vessels [11]. A complete surgical resection of the tumor from the vagus nerve is the goal of the treatment with attempt to preserve the vagal pathway [7]. If the complete resection of the tumor with preservation of the vagus nerve was impossible, the best choice in this case is to reset the involved segment and end to end anastomosis [8]. The most common complication postoperatively is hoarseness, so preoperative focal cord mobility assessment is necessary [5]. In preoperatively assessment, vocal cord paralysis has been reported in 12% of cases [5,8]. Whereas postoperatively, vocal cord paralysis was reported in 85% of cases [8]. In case of vocal cord paralysis postoperatively, aggressive voice therapy should start as soon as possible after the surgery to compensate the vocal cord [8]. Other possible complications are dysphagia, dysphonia, Horner's syndrome, facial myotonia, hypoglossal palsy, and facial palsy [8]. We have presented a rare case of vagal schwannoma in an adolescent male with a few previously published cases in the literature [5].

Competing Interests

The authors declare that there is no conflict of interests regarding the publication of this paper

References

1. Kang GC. Extracranial non-vestibular head and neck schwannomas: a ten-year experience. *Ann Acad Med Singapore*. 2007;36(4):233-238.
2. Quick CRJ, Reed JB, Harper SJF, Saeb-parsy K, Burkitt HJ. *Essential surgery: problems, diagnosis, and management*. (5th ed.). Scotland: Elsevier. 2014.
3. Chiofalo MG, Longo F, Marone U, Franco R, Petrillo A and Pezzullo L. Cervical vagal schwannoma. A case report. *Acta Otorhinolaryngol Ital*. 2009;29(1): 33-35.
4. Cavallaro G, Pattaro G, Iorio O, Avallone M, Silecchia G. A literature review on surgery for cervical vagal schwannomas. *World Journal of Surgical Oncology*. 2015;13:130. doi:10.1186/s12957-015-0541-6
5. Sreevatsa M, Srinivasarao R. Three Cases of Vagal Nerve Schwannoma and Review of Literature. *Indian Journal of Otolaryngology and Head & Neck Surgery*. 2011;63(4):310-312. doi:10.1007/s12070-011-0220-z
6. Kamath P. Vagal Schwannoma: A Rare Parapharyngeal Tumour. *JOURNAL OF CLINICAL AND DIAGNOSTIC RESEARCH*. 2016;10(4):MD03-MD04. doi:10.7860/JCDR/2016/16391.7588
7. Singh ID. Cervical Vagal Schwannoma Presenting as Paroxysmal Cough-A Case Report. *Journal of Otolaryngology-ENT Research*. 2017;7(3). doi:10.15406/joentr.2017.07.00206
8. Giancarlo Andrea D, Isabella Famà, Roberto Morello, Roberto Becelli. Cervical Vagal Schwannoma, Review of all Reported Cases and Our Reports. *Int J Neurol Brain Disorders*. 2016;3(2):1-6.
9. Yong, D, Hailani I, Bin Mohamad YM. Cervical Vagal Schwannoma. *Philippine Journal of Otolaryngology Head and Neck Surgery*. 2010;25(2).
10. Anil G, Tan T. CT and MRI Evaluation of Nerve Sheath Tumors of the Cervical Vagus Nerve. *American Journal of Roentgenology*. 2011;197(1):195-201. doi:10.2214/AJR.10.5734.
11. Anvekar, D. Salt and Pepper sign [Internet]. *Neuroradiology cases*. 2017