



Surgical Resection of Vagal Schwannoma in a Young Female

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Abstract

Background: Paraganglioma of head and neck is a rare tumor and vagal schwannoma is even rarer. The majority of patient with vagal schwannoma presents with lateral cervical neck mass. Typically, it is a slow growing, painless and mobile tumor but it has high tendency to become malignant compare to other subtypes of paraganglioma. The choice of treatment includes surgery, radiation or observation and it depends on the characteristic of tumor as well as patient's preference.

Case Description: A 22-year old lady presented with history of a right neck mass for 5-months duration. Clinical examination revealed a mass at level II neck region which measures 3.0 cm × 2.0 cm and it was mobile, non-pulsatile and had smooth surface. CT scan and angiogram showed that the mass arises between carotid artery and vagal nerve and it was a highly vascular lesion. A CT scan-guided biopsy performed but complicated with neck hematoma and patient developed hoarseness. On follow up, her hoarseness persists and her tissue biopsy came back as schwannoma. She was counselled regarding surgery versus radiation for her treatment and she agreeable for surgery. Hence, surgical excision was performed and intraoperatively it showed that the mass arose from the vagal nerve.

Conclusion: Vagal schwannoma is a rare paraganglioma of head and neck. Patient's symptom as in this case, the complaint of hoarseness can give clue to the origin of the lesion and meticulous extirpation of the mass should be practiced in order to avoid inadvertent injury to intimate important neurovascular structures.

Keywords: Vagal nerve; Schwannomas; Paraganglioma; Head and neck; Excision

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Introduction

Paraganglioma of head and neck is a rare tumor and accounting for only 1% of all head and neck tumor. There are several types of paraganglioma that occurs mostly in the neck which includes glomus tumor, carotid body tumor and schwannoma. The majority of patient however is asymptomatic as the tumor is benign and has indolent growth. Clinical presentation depends on types and locations of the tumor and the surrounding neurovascular structural involvement. Temporal bone paragangliomas or glomus tympanicum tumor normally originates in the middle ear, either from the Jacobson's or Arnold's nerves and characteristically shows bluish mass behind tympanic membrane. The patient glomus tympanicum tumor may present with complaint of hearing loss, tinnitus and disequilibrium [1]. On the other hand, carotid body tumor grows within bifurcation of carotid artery and generally present with a painless, mobile and slow growing mass over the neck region. This tumor type is commonly pulsatile and transmits the bruit.

Schwannomas of the neck is rare type of paraganglioma and patient may present with neck swelling. If the schwannomas arise directly from the vagal nerve, the patient may be complaining of hoarseness or sometimes cough on palpation of the neck mass. In addition, with sympathetic nerves schwannoma, clinical examination will reveal Horner's syndrome which is characterized by ptosis, mydriasis and pupillary constriction or meiosis on the affected side.

The standard management approach for paraganglioma includes surgery, radiation, or watchful observation especially if it is asymptomatic. The symptomatic lesions may justify surgical resection if the expertise is available and consented by patient. Of note, current trends have shifted to a more conservative approach for managing paragangliomas in view of its benign behavior with indolent growth and causing minimal symptoms. Head and neck paragangliomas may occur as a sporadic or hereditary tumor. If it occurs as a hereditary trait, the paraganglioma may occur in association with sympathetic catecholamines-secreting paragangliomas; located in abdomen or in the chest and they



Figure 1: Clinical examination revealed a mass at right level II of the neck which measures 3.0 cm x 2.0 cm, rounded, mobile, non-pulsatile with smooth surface.

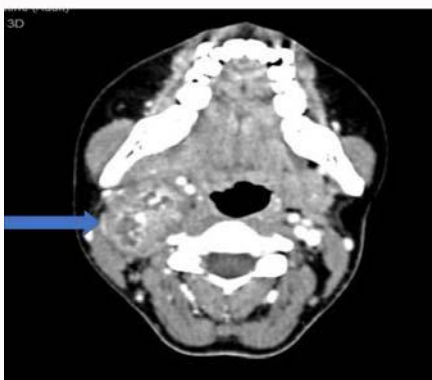


Figure 2: CT scan of the neck showed a heterogenous, well encapsulated tumor which located between the carotid artery and the vagal nerve.

may occur as multiple tumors [2].

Case Presentation

A 22-year old Malay girl presented in early 2013 with a small right cervical neck swelling. Investigation performed at that time revealed a mass measuring 3.0 cm x 4.0 cm and it was a highly vascular tumor on the angiogram. Patient was offered for treatment with radiation or surgery, however patient refused and defaulted treatment. In 2015, she represented with increasing neck swelling associated with mild pain. There was no other significant complaint. Clinical examination revealed a mass at level II neck node measuring 3.0 cm x 2.0 cm, mobile, non-pulsatile, and with smooth surface (Figure 1). After discussion with the interventional radiologist, patient was decided for tissue biopsy under CT scan guided, and possibly with embolization in view of its vascularity. Patient underwent the biopsy under CT scan guided but post procedure patient develops swelling at the neck and hoarseness. On repeat clinical examination, post procedure, there was hematoma at the neck region from the biopsy site. This hematoma had caused compression on the vagal nerve and caused recurrent laryngeal nerve paralysis with resultant hoarseness. A few days later, patient was improved and was discharged home well and on subsequent follow up, it was noted that the neck hematoma has subsided but hoarseness persists. Endolaryngeal examination revealed right vocal cord palsy with minimal phonatory gap. The tissue biopsy results came back as schwannomas. Repeated CT scan showed the tumor was located between the carotid artery and the vagal nerve and it is a well-defined tumor with good demarcation

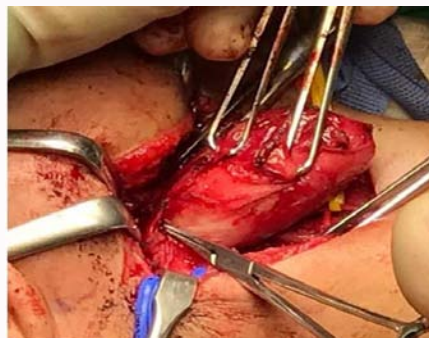


Figure 3: Intraoperatively, the mass was observed arising directly from the vagal nerve, in between the carotid artery and internal jugular vein.

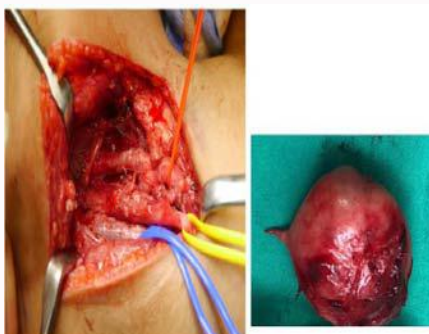


Figure 4: The mass was removed in total and measures 5.0 cm x 4.0 cm, leaving the vagal nerve intact (identified with yellow vessel loupe) with carotid artery and internal jugular vein (identified with red and blue vessel loupe respectively).

from the surrounding tissue (Figure 2). After lengthy discussing with the patient and her family, the patient was agreeable for surgery.

Intraoperatively, patient was in supine position with slight neck extension and faced to contra lateral side and the lesion and the surgical anatomy landmarks were drawn, taking account preservation of the marginal mandibular nerve and expected cosmesis and ease of access to the carotid sheath. The skin incision performed and the skin flap was raised superiorly and inferiorly. The anterior border of the sternocleidomastoid muscle is skeletonized and exposing the mass deep to the sternomastoid. Continues dissection was performed and later, the carotid artery was identified and retracted laterally. The internal jugular vein was visualized deep and medial to the mass and was slowly retracted medially. The vagal nerve is difficult to identify initially but after deeper and careful dissection, the mass was observed arising directly from the perineurium of the vagal nerve (Figure 3), in between the carotid artery and internal jugular vein. With gentle and meticulous dissection, the vagal nerve is separated from the mass. The mass was removed in total and measured 5.0 cm x 4.0 cm (Figure 4), leaving the vagal nerve intact. The homeostasis was secured, and the wound closed in 2 layers with small drain secured in situ. The patient was discharged home uneventfully except for her hoarseness. The post-operative tissue biopsy result came back as vagal schwannomas and patient was doing well on the last follow up. Repeated laryngoscopy showed her right vocal cord remain paresis with partial compensation and minimal phonatory gap. Her voice however, improved and she was pleased with the surgery.

Discussion

Paragangliomas is a benign tumor, and in the head and neck

region its constitute about 1% of all tumors. In most cases, it is a slow growing tumor and rarely causes severe symptoms. Details clinical presentation however depends on multiple factors such as the size of the tumor and its location. The standard management for head and neck paraganglioma includes close observation, surgical excision, radiation or stereotactic radio surgery and in selected cases it requires a combination of treatment. Both surgery and radiation may provide a good control of local tumors but with different complications which depends on the tumor extent and location. It has been documented that this paraganglioma may be associated with malignant transformation especially in selected long standing cases. On the other hand, Gilbo et al. [1] divided the paraganglioma into benign and malignant type [1]. They further stated that the malignant variant of paragangliomas is not based on the histological diagnosis, but determined by its clinical features, development of metastases and complaint of pain.

The presentation of paragangliomas can be subtle. Interestingly, however multiple symptoms have been reported in literatures ranging from hoarseness, sensorineural hearing loss, Horner's syndrome, neck mass and hypoglossal nerve palsy. The commonest symptom however is depending on the location and size of the tumor. In this case, the patient had a neck mass that was progressively increase in size and hoarseness that develops after tissue biopsy strongly suggested that the mass was in close proximity with the vagal nerve. If surgical excision decided for her treatment, the morbidity of the surgery was less as anticipated hoarseness already present prior to the surgery. The aim of surgery however is to preserve the integrity of the vagal nerve and its function. Imperatively, in this case her hoarseness was improving gradually postoperatively with no other complaints such as history of aspiration.

According to Smith et al, vagal paragangliomas were the biggest tumor in their study cohort with size of 5.3 cm (+/-1.9 cm). They further mentioned that most common mode of treatment is surgical excision with preoperative embolization. The resultant complications include complete unilateral vagal nerve paralysis together with additional glossopharyngeal, hypoglossal nerve and cervical sympathetic chain disturbances [3]. Heyes et al. [4], suggested that surgical excision is the standard treatment of choice for most vagal schwannomas but they stressed that nowadays, the contemporary management evolves toward a conservative modality due to high morbidity of the surgery. They further stated that vagal schwannomas resection almost always requires vagal nerve scarification with resultant speech, swallow and sensory deficits [4].

Watchful waiting with serial imaging also has been successfully employed. Generally, most of lesions remain radiologically stable in the majority of cases. In few selected cases of schwannomas, there was some development and progression of neuropathy observed. Watchful waiting is especially valuable in asymptomatic older patients. This watchful waiting policy has its advantageous of preventing the surgery and its potential complications [5,6]. This is specifically true if the mass is small and does not cause any symptoms. The subsequent treatment of schwannomas also depends very much on the final tissue diagnosis of the tumor. The surgical excision is advocates in any symptomatic cases with progressive disease as well as if the suspicion of malignancy is high in selected cases. The downside is that if the tumor even though it is small, found to be a malignant tumor and has propensity for distant metastases. This crucial point justified why in this case we proceed with tissue biopsy even though we know it is a highly vascular tumor. If the biopsy result came back

as malignant tumor, the treatment will be different where radiation should be incorporated in her overall treatment plan.

Numerous surgical approaches have been documented in the literatures and mainly depend on the location of the tumor and its surrounding tissue involvement. For the neck paraganglioma, the traditional transcervical skin collar excision is opted with supratyma flap. This will adequately expose the tumor after meticulous dissection. The identification and preservation of the carotid artery and the internal jugular vein as well as the vagal nerve is paramount. In this case, the carotid artery and internal jugular vein were retracted medially and laterally respectively to get better surgical plane surround the tumor. The tumor arises from the perineurium of the vagal nerve as it is closely adherent to the vagal nerve. The vagal nerves together with hypoglossal nerve were preserved.

The other approach of the tumor can be transtemporal, transoral, transmandibular and combination of these approaches. The transmandibular approach is suitable for deep seated paraganglioma especially in the parapharyngeal spaces. In selected cases, the mandibulotomy has to be performed and the facial nerve need to be identified, preserve and mobilized laterally. Tran temporal approach is used for glomus tympanicum and glomus jugulare tumors and can be combined with other approaches to better suit the details characteristic of the tumor. The approach for glomus jugulare tumor sometimes need to be combined with the neurosurgical approaches such as bicoronal craniotomy in order to excise the tumor in monobloc fashion [6,7]. The complications from surgery can be multiple and mainly depends on the experiences and wisdom of the attending surgeons and the availability of the multidisciplinary team expertise are vital to combat unwanted surgical morbidities. These complications can be minimized in the expert hands and the outcomes of postoperative can be improved. This is imperative for the patient's quality of life as in this case patient is still young and there is a long path ahead so preserving the voice and the aesthetic for her is paramount.

The other treatment modality for head and neck paraganglioma includes fractionated radiation and stereotactic radio surgery. According to Smith et al., this treatment approach was most commonly employed for the jugular and vagal paragangliomas. In their study they found out that the radiation produces excellent tumor control for 2-119 months with no major treatment complications [2]. This treatment types are best suited large or multifocal head and neck paragangliomas who have high likelihood of surgical morbidity to last for cranial nerves and those who are contraindicated for surgery due to medical comorbidities. Gilbo et al. [1], stated that fractionated radiation dose required to treat benign paragangliomas is 45 Gray at 1.8 Gray per once daily fraction. Higher dose provides no improvement of local control and results in an increase in complications [1].

Most genetic studies on paraganglioma patients have highlighted the germline mutations in the RET, NFI and SDH mutations. According to Piccini et al. [2], the gene mutations responsible for head and neck paraganglioma occurs mostly in genes encoding the subunits of succinate dehydrogenase or mitochondrial complex II which includes SDHD which located on 11q2 chromosome [2]. They revealed that a positive family history, presence of multiple head and neck paragangliomas and association with sporadic paragangliomas were invariably characterized by a germ line mutation. The SDH mutation has been associated with clinical paraganglioma syndrome.

Gilbo et al. [1] documented that individual who are more likely to have SDH mutations are those who present with family history of paragangliomas, a previous pheochromocytoma, multiple head and neck paragangliomas, age less than 40 years old and a male [1]. Genetic testing for hereditary head and neck paragangliomas is important as it will allow screening those family members and individuals at high risk as well as for mitigating life-long surveillance protocol.

Conclusion

Vagal schwannomas at the cervical region is an important clinical entity. Treatment can either be a surgery, observation or radiation and it depends on tumor characterization and patient's preference. The primary objective of any treatment strategy is to alleviate the patient's symptoms and to optimize post-operative outcomes as well as prognosis. Selected cases of schwannoma, can be offered surgical excision to reduce the symptoms from expanding mass, improves the aesthetic as well as to ensure better quality of life by avoiding the radiation and its sequel.

References

1. Gilbo P, Morris CG, Amdur RJ, Werning JW, Dziegielewski PT, Kirwan J, et al. Radiotherapy for benign head and neck paragangliomas: a 45-year experience. *Cancer*. 2014;120(23):3738-43.
2. Piccini V, Rapizzi E, Bacca A, Di Trapani G, Pulli R, Giache V, et al. Head and Neck Paragangliomas: genetic spectrum and clinical variability in 79 consecutive patients. *Endocr Relat Cancer*. 2012;19(2):149-55.
3. Smith JD, Harvey RN, Darr OA, Prince ME, Bradford CR, Wolf GT, et al. Head and Neck Paragangliomas: A Two decades institutional experience and algorithm for management. *Laryngoscope Investig Otolaryngol*. 2017;2(6):380-9.
4. Heyes R, Taki N, O'Leary MA. Vagal paraganglioma presenting as a neck mass associated with cough on palpation. *Case Rep Otolaryngology*. 2017;7603814.
5. Moore MG, Netterville JL, Mendenhall WM, Isaacson B, Nussenbaum B. Head and Neck Paragangliomas: An update on evaluation and management. *Otolaryngol Head & Neck*. 2016;154(4):597-605.
6. Szymanska A, Szymanski M, Czekajska-Chehab E, Golabek W, Szezerbo-trojanowska M. Diagnosis and management of multiple paragangliomas of the head and neck. *Eur Arch Otorhinolaryngol*. 2015;272(8):1991-9.
7. Tareb D, Kaliski A, Boedecker CC, Martucci V, Fojo T, Adler JR, et al. Current approaches and recent developments in the management of head and neck paragangliomas. *Endoc Rev*. 2014;35(5):795-819.