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TITLE: Neurofibroma of the cervical part of the vagus nerve: A case report

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ABSTRACT

Introduction
Neurofibroma of the vagus nerve in the cervical region is an extremely uncommon benign tumour. Only ten reported cases were found in the literature. It is slow growing and most are asymptomatic neck lumps. Preoperative imaging aids in arriving at a differential diagnosis and planning the surgery. Complete surgical excision is the standard of care.

Case Report
A 39 year old female presented to the General Surgical clinic with a painless lump on the Right side of the neck for twelve months duration with recent rapid enlargement. Past medical history was unremarkable. She did not have a family history of neurofibromatosis. Examination revealed a 4 x 3.5cm lump in the upper neck with well-defined margins and smooth surface. Contrast enhanced CT scan confirmed a 3.6 x3.2 x6.9cm hypodence mass with no contrast enhancement. The lesion was in-between Right internal and external carotid artery and extending to the base of the skull. Right internal jugular vein (R/IJV) was displaced anteriorly. Right common carotid artery(R/CCA) was stretched.

The patient underwent exploration of the neck under general anaesthesia with loupe magnification. The gross pathology showed a greyish homogenous cut surface of a 55x40mm tumour. Pathological examination confirmed the diagnosis of neurofibroma of the vagus nerve.

Conclusion
Vagus nerve neurofibromas are rare nerve sheath tumours. Gold-standard treatment of symptomatic benign vagus nerve mass is an attempt at GTR with minimal loss of nerve function.

Keywords: Neurofibroma, vagus nerve, tumour
TITLE: Neurofibroma of the cervical part of the vagus nerve, A case report

INTRODUCTION
Neurofibroma of the vagus nerve in the cervical region is an extremely uncommon benign tumour [1]. Only ten reported cases were found in the literature [2]. It is slow growing and most are asymptomatic neck lumps [3]. Preoperative imaging aids in arriving at a differential diagnosis and planning the surgery. Complete surgical excision is the standard of care. Here we present a case of sporadic neurofibroma of the vagus nerve in the cervical region of a 39 year old lady. She underwent successful surgery without disability such as vocal cord palsy.

CASE REPORT
A 39 year old female presented to the General Surgical clinic with a painless lump on the Right side of the neck for twelve months duration with recent rapid enlargement. Past medical history was unremarkable. She did not have a family history of neurofibromatosis. Examination revealed a 4 x 3.5cm lump in the upper neck with well-defined margins and smooth surface. Upon palpation paroxysmal cough was not elicited.

Ultrasound (US) of the neck showed a well-defined solid soft tissue mass with internal vascularity. Contrast enhanced CT scan confirmed a 3.6 x3.2 x6.9cm hypodence mass with no contrast enhancement. The lesion was in-between Right internal and external carotid artery and extending to the base of the skull. Right internal jugular vein (R/IJV) was displaced anteriorly. Right common carotid artery (R/CCA) was stretched (Figure 1).

The FNAC of the mass was inconclusive showing only stromal tissue fragments. The patient underwent exploration of the neck under general anaesthesia with loupe magnification. A vertical incision is made along the anterior border of the sternocleidomastoid muscle achieving vascular control. An ovoid whitish tumour was identified in between the R/IJV and the carotid artery displacing them. Both the superior and inferior ends of the mass appeared in continuity with the vagus nerve stretching its nerve fibres. Plane of the tumour was approached splitting the nerve
fibres in the longitudinal direction. Tumour was completely excised preserving the
continuity of the vagus nerve (Figure 2).

The gross pathology showed a greyish homogenous cut surface of a 55x50mm
tumour. Pathological examination confirmed the diagnosis of neurofibroma of the
vagus nerve.

Microscopy showed loosely arranged spindle cells containing slender wavy nuclei
with pointed edges. Scattered cells showed enlarged hyperchromatic nuclei
resembling degeneration. Mitoses were not increased (Figure 1).

Patient recovered well without hoarseness and discharged home in the second
postoperative day. She was followed-up in the clinic with no major complications.

DISCUSSION

Neurofibromas of the vagus nerve are an extremely rare peripheral nerve sheath
tumour. There occurrence in the cervical region is even uncommon. There are only
ten reported cases of such lesions found in the literature [2]. Majority of neoplasms
of vagus nerve are schwannomas [4,5].

Neurofibromas of the vagus nerve are slow growing peripheral nerve sheath tumours
[3,6]. Generally they are asymptomatic. Patients present with a painless neck lump
with no significant disability. Some case studies have documented dysphagia and
cough with percussion of the mass [4]. Commonest age range is 20-40 years with no
sex predisposition [12]. There may be an association (60%) with neurofibromatosis
type 1 [8, 2].

Growth takes the fusiform shape surrounded by a pseudo-capsule with the stretched
neural tissue. Most are benign with a malignant counterpart less frequently.

Preoperative imaging with MRI is preferred, to assess the tumour characteristics and
extension and arriving at a differential diagnosis. Ultrasound scan and CT are also
valuable imaging modalities especially in limited resource setting. Optimal surgical
approach and planning of surgery is made according the findings.

Cytological assessment with FNAC is non-specific. Biopsy is discouraged due to risk
of damaging the surrounding structures, bleeding and because of its effects on future
definitive surgery.
Gross total resection is the definitive treatment (GTR) [5]. Neurofibromas prove to be most difficult nerve sheath tumours to achieve GTR. Removal of the tumour by internal decompression is preferred as it preserves the function and minimizes the damage to closely packed structures in the vicinity [10]. If the neurofibroma is removed enbloc, microsurgical repair of the nerve is indicated with a nerve graft [13]. Following the excision complications such as vocal cord palsy, dysphagia and arrhythmia could occur. Donner et al previously reported a 16% decrease in motor function using the intra-capsular enucleation technique. Among them Up to 10% developed pain syndromes [8]. Green et al reported a post-operative complications like occurrence of dysphagia (40%) and aspiration (46%) after vagus nerve tumour surgery [7]. Gilmer-Hill and Kline stated that nearly all patients who have vagus nerve tumour resections will develop transient hoarseness postoperatively [14]. Our patient did not develop any of these major complications.

Patients should be followed-up for local recurrence because of it’s a higher possibility [12].

CONCLUSION

Vagus nerve neurofibromas are rare nerve sheath tumours. Gold-standard treatment of symptomatic benign vagus nerve mass is an attempt at GTR with minimal loss of nerve function. Postoperative hoarseness and vocal cord paralysis can be avoided with meticulous surgical technique.

CONFLICT OF INTEREST

None of the authors have any conflicts of interest

AUTHOR’S CONTRIBUTIONS

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Group 1 - Substantial contributions to conception and design, acquisition of data, or analysis and interpretation of data

Group 2 – Drafting the article

Group 3 - Final approval of version to be published
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REFERENCES


FIGURE LEGENDS

Figure 1: CT view of right sided hypodence mass with no contrast enhancement

Figure 2: Ovoid tumour in-between the R/IJV and the carotid artery

Figure 3: Loosely arranged spindle cells
FIGURES

Figure 1: CT view of right sided hypodence mass with no contrast enhancement

Figure 2: Ovoid tumour in-between the R/IJV and the carotid artery
Figure 3: Loosely arranged spindle cells