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Neurofibroma at an Aberrant Location - A Case Report

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ABSTRACT

Background: Neurofibromas are benign tumors of peripheral nerve sheath and are uncommon in cervical part of vagus nerve.

A 35-year female presented with nontender swelling on the left side of the neck since 2 months with no family history. Clinical diagnosis was made as? carotid body tumor/parapharyngeal mass.

CT neck showed well defined hypodense oval mass in the left upper neck in the carotid space reported as features suggestive of schwannoma. Post-operative diagnosis of left vagal schwannoma was made and sent for histopathological examination.

Gross morphology was well circumscribed grey white mass measuring 6x4x3cms with gelatinous cut surface. On microscopy it was neoplasm composed of spindle cells with poorly defined cell borders having buckled nuclei with pale eosinophilic cytoplasm. These cells are arranged loosely in the background of collagenous matrix. No evidence of atypia or malignancy noted. Features consistent with Neurofibroma.

Conclusion: Histopathological confirmation is essential for the diagnosis of neurofibroma in cervical part of vagus nerve as it is an uncommon site for the lesion to develop.

Key words: Neurofibroma, Cervical part of vagus nerve, Aberrant site.

INTRODUCTION

Neurofibromatosis was first described by the physician Mark Akenside in 1768.[1] These are benign peripheral nerve sheath tumor with classic identifiable features including the presence of a neuronal component comprising transformed Schwann cells and a non-neoplastic fibrous component that includes fibroblasts.

Neurofibromas have extremely low risk for malignant transformation and most commonly occurs as part of generalized syndrome of neurofibromatosis.[2] However they are not associated with neurofibromatosis type I when they occur as solitary lesion.[3]

90% of cases occur sporadically while remaining are associated with Neurofibroma type I/II following a mutation/deletion in NF1 gene. Neurofibroma of cervical part of vagus nerve are very uncommon. They are slow growing and present as asymptomatic neck masses.[4]

CASE REPORT

A 35-year female presented with nontender swelling on the left side of the neck since 2 months which was insidious in onset and progressive in nature. There was no history of NF in the family.

On examination- Mass measured 4x3cms and firm in consistency. Clinical diagnosis was made as? carotid body tumor/parapharyngeal mass.

CT neck showed well defined hypodense oval mass in the left upper neck in the carotid space measuring 65x51x26mm reported as features suggestive of schwannoma.

The mass in toto was excised under general anesthesia. During the procedure thinning

and splaying of carotid vessels were noted and post-operative diagnosis of left vagal schwannoma was made and sent for histopathological examination.

Gross morphology showed well circumscribed grey white mass measuring 6x4x3cms with gelatinous cut surface (figure 1).

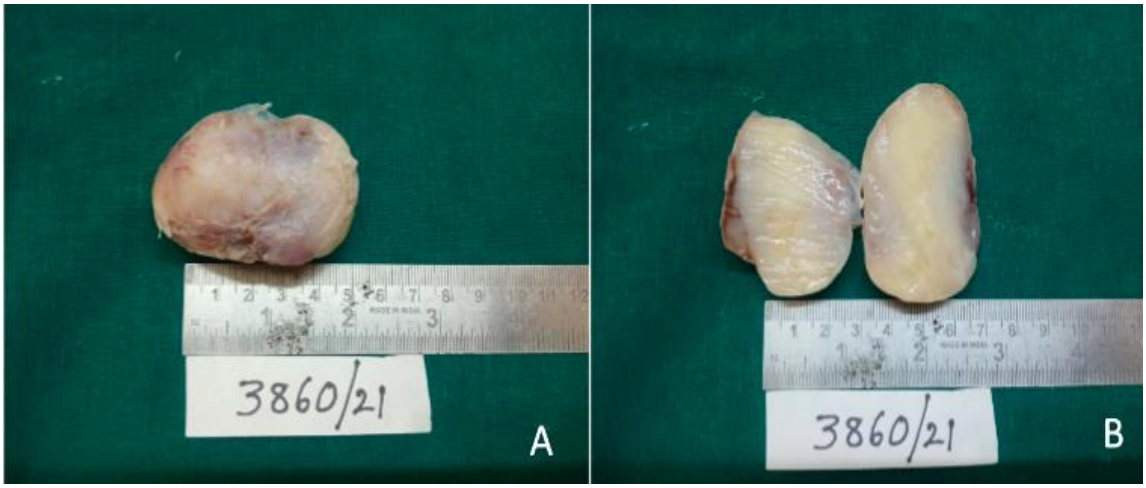

Figure 1: A: External surface, B: Cut section showing gelatinous areas.

Microscopy showed neoplasm composed of spindle cells with poorly defined cell borders having buckled nuclei with pale eosinophilic cytoplasm. These cells are arranged loosely in the background of collagenous matrix. No evidence of atypia or malignancy noted (figure 2).

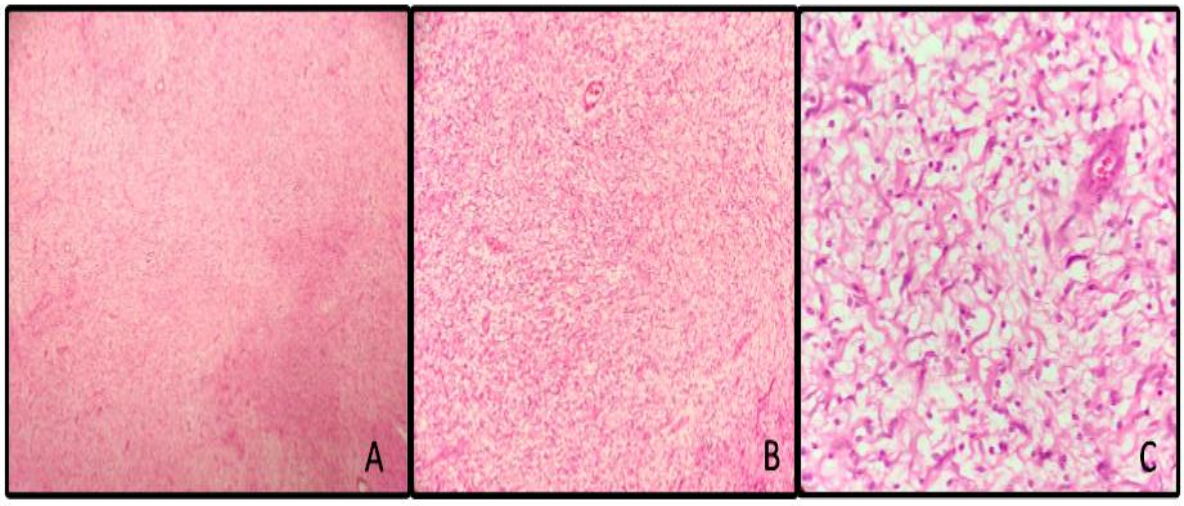

Figure 2: Microscopy A)4X, B)10X, C)40X showing spindle cells with buckled nuclei in a collagenous matrix.

DISCUSSION

Neurofibromas are slow growing benign tumors of peripheral nerve sheath and are extremely rare in cervical part of vagus nerve.

Clinically neurofibromas can occur as subcutaneous or plexiform varieties. Subcutaneous variety accounts for 95% of cases and is seen at peripheral nerve endings. Plexiform variety accounts for 5% of cases and is seen near the nerve roots. Plexiform variety has high potential for malignant transformation.[5]

Neurofibromas have to be differentiated from schwannomas and carotid body tumors which are more common in this location.[6] Solitary neurofibromas are unencapsulated and ill circumscribed lesions. Histologically, consists of elongated spindle cells with poorly defined, pale eosinophilic cytoplasm and tapering, wavy or buckled nuclei admixed with small nerve fibers and frequent mast cells. Stroma is fibro myxoid, mucinous or hyalinized.[7]

Matejcik V et al reported one case of neurofibroma of cervical part of vagus nerve which was the single case reported in their experience of 22 years. They also found only 9 described cases of surgical treatment of the neurofibroma of the vagus nerve in the neck region in literature available till 2007.[8]

Carotid splaying, also known as Lyre sign is not a common feature of neurofibromas. This finding is noted in our case and also reported by Itawi SA et al.[9]

Complete surgical excision is the gold standard treatment in case of solitary neurofibroma and same was done in our case.

CONCLUSION

Cervical part of the vagus nerve is an uncommon site for solitary neurofibromas. Histopathological confirmation is must to rule out other differential diagnoses which can mislead clinically.

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Ethical consideration: As the study required only routine histopathology slides with no additional financial requirements, and consent being taken from every patient at the time of hospital admission regarding utilization of data for research purpose, the protocol did not need formal ethical clearance from review board.

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