Case Report

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Schwannoma neck: a diagnostic dilemma

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ABSTRACT

Schwannomas are less common benign slow growing tumors originating from Schwann cells. In the head and neck region, schwannomas arise most commonly from the vagus nerve or the sympathetic chain. We present this case as the location of schwannoma is extremely rare and due to the diagnostic difficulties it posed. A 48 year old male presented with right neck swelling and breathing difficulty to our OPD. Patient underwent ultrasonogram of neck, MRI neck and Fine needle aspiration cytology (FNAC) of the lesion. Each of the investigations suggested different pathology which made the diagnosis challenging. During surgery, the lesion was found to arise from right recurrent laryngeal nerve. After excision of the lesion, the patient developed hoarse voice and the pathological examination revealed schwannoma. Schwannomas that originate from Schwann cells can affect any part of the body. MRI, CT, USG and FNAC have been suggested in the literature for diagnosing the lesion. Trucut biopsy should be considered in situations where FNAC becomes inconclusive. Surgical excision is the treatment of choice. Histologically, five variants of schwannomas have been described in the literature namely common, plexiform, cellular, epithelioid and ancient schwannoma. To conclude, schwannoma arising from RLN which masqueraded as a thyroid swelling is a rare entity. The diagnostic modalities suggested in the literature were unable to pin point the diagnosis. Once, FNAC shows an inadequate specimen, a trucut biopsy should be considered as the next investigation modality.

Keywords: Schwannoma, Recurrent laryngeal nerve, Thyroid gland

INTRODUCTION

Schwannomas are less common benign slow growing tumors originating from Schwann cells. It was first described by Verocay in 1908, and the first reported case was by Delaney and Fry in 1964.¹ About 25–40% of cases occur in the head and neck region.² In the head and neck region, schwannomas arise most commonly from the vagus nerve or the sympathetic chain.³ Signs and symptoms of head and neck schwannoma arising from recurrent laryngeal nerve (RLN) is a rare entity which can lead to misdiagnosis as thyroid nodule. In this case, we describe extremely rare location for schwannoma, which posed diagnostic difficulties, and possible role of trucut biopsy in diagnosing the condition.

CASE REPORT

A 48 year old male came with complaints of neck swelling for 9 months and breathing difficulty for 7 was months which gradually progressive. On examination, there was a swelling $\sim 5 \times 4$ cms in the anterior aspect of neck more towards the right side extending from the midline medially to the posterior border of the right sternocleidomastoid muscle laterally. Superiorly it was 5 cms below the chin and the inferior border could be palpable when the swelling moved upwards while swallowing. There was no movement on protrusion of the tongue. Indirect laryngoscopy showed that both vocal cordswere mobile. Initially, a clinical diagnosis of right thyroid nodule was made. USG neck done from two outside hospitals suggested a right hypoechoic thyroid nodule measuring $\sim 56.9 \times 29.8$ mm².

Fine needle aspiration cytology (FNAC) of the nodule done from both the places was non diagnostic. We obtained an endocrinologist opinion and the patient was sent for a repeat real time USG neck. USG suggested schwannoma or neurilemmoma (Figure 1). To get a clear picture, we obtained MRI neck which suggested that it could be a parathyroid adenoma (Figure 2), but serum PTH was 65.4 pg/ml and serum calcium- 9 mg/dl; both were normal. A rereporting of the MRI neck suggested it could be a thyroid nodule or a schwannoma. The differential diagnosis after the extensive investigations was right thyroid nodule or a schwannoma. To relieve patient's symptoms, we took the patient for surgery. Intraoperatively, we found $\sim 6 \times 4$ cms well encapsulated lesion which was easy to dissect, lying posterior to right lobe of thyroid and attached to the posterior fascia of thyroid gland. The lesion was extending from below cricothyroid muscle to above the clavicle. The lesion was found to be arising from right RLN (Figure 3), we preserved the nerve integrity by performing enucleation. We also identified the vagus nerve which was found to be normal. Post operatively, patient developed hoarse voice and clinical examination confirmed right vocal cord palsy. Histopathological examination (HPE) showed encapsulated neoplasm composed of fascicles of spindle shaped cells exhibiting nuclear palisading (Antoni A area) along with loose reticular areas (Antoni B area) suggestive of schwannoma (Figure 4).

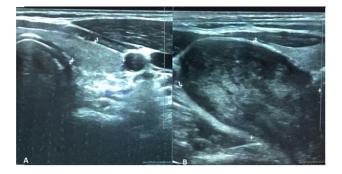


Figure 1: (A) Normal left lobe of thyroid, (B) Schwannoma lying posterior to right lobe of thyroid.

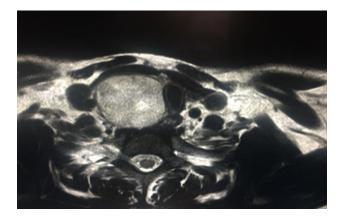


Figure 2: T2W MRI showing heterogeneously hyperintense lesion posterior to right lobe of thyroid.

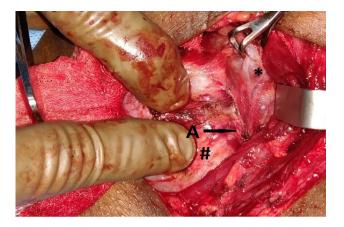


Figure 3: Lesion (#) arising from recurrent laryngeal nerve (A), (*) right lobe of thyroid marked.

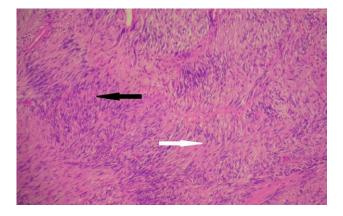


Figure 4: HPE of lesion, black arrow indicating Antoni A area, white arrow indicating Antoni B area.

DISCUSSION

Schwannomas are benign neurogenic tumors arising from Schwann cells. Schwann cells are derived from the neural tube and are part of the glial cells. The main function of Schwann cell is to form the myelin sheath which acts as electrical insulator and thereby contributing for better conduction of nerve stimuli. Schwannomas can arise from any cranial or peripheral nerve in any part of the body; in the head and neck area, it constitutes 25-40%.² Usually, notable signs or symptoms are not present in the early stages of the disease. A solitary, slow growing mass in the neck is the most common symptom. It is believed to be a slow growing tumor, Zhang et al and de Araujo et al reported annual rates of tumor growth of 2.75 mm and 3 mm respectively. Malignant change in schwannomas of the head and neck is rare, with a prevalence of 8–13.9%.⁴

MRI, CT, USG and FNAC have been suggested in the literature for diagnosing the lesion.⁵ USG can help to differentiate the lesion from thyroid in the hands of experienced sonologist as seen in our case. MRI is reliable imaging to capture the tumor, its capsule and the nerve from which the tumor arises.⁶ In 1996, Furukawa et al undertook imaging studies on nine patients with schwannomas and suggested their neurological origin

prior to surgery and they were correct in all cases. In 2007, the accuracy of the preoperative diagnosis made by Saito et al in 12 patients with schwannomas was 83%.⁴ Another preoperative imaging study done by Kim et al in 7 patients suggested that schwannoma arising from sympathetic chain displaces both Internal jugular vein (IJV) and carotid anteriorly meanwhile, schwannomas originating from Vagus displaces IJV laterally and carotid medially, but in our case the lesion has pushed both carotid and IJV laterally.³ Although FNAC has been suggested for cytological diagnosis, a review literature suggests that it is rarely conclusive.^{5,7-9} Trucut biopsy might play a role in tissue diagnosis ahead of planning the surgery and thereby to consent the patient regarding the morbidity and prognosis.

Surgical excision is the treatment of choice; it is sometimes possible to preserve the nerve function by carefully performing microsurgical nerve sparing surgery along with nerve monitoring.¹⁰ In our case, even though we preserved the nerve integrity, the patient ended up with right vocal cord palsy. Histologically, five variants of schwannomas have been described in the literature namely common, plexiform, cellular, epithelioid and ancient schwannoma.² On histopathological examination, it is composed of an intimate mixture of spindle cells forming highly cellular Antoni A areas and less cellular myxoid Antoni B areas.⁵ Immunohistochemistry is further more confirmatory which are positive for S100 and Vimentin and negative for Desmin and SMA.¹

CONCLUSION

To conclude, schwannoma arising from RLN which masqueraded as a thyroid swelling is a rare entity. The diagnostic modalities suggested in the literature were unable to pin point the diagnosis. Once, FNAC shows an inadequate specimen, a trucut biopsy should be considered as the next investigation modality.

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