# **Case Report**

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# Ancient schwannoma of cervical sympathetic chain masquerading as carotid body tumour

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### **ABSTRACT**

Schwannomas are benign tumours arising from the nerve sheath cell. 25-40% of schwannomas are found in head and neck. Ancient schwannona of cervical sympathetic chain is very entity. It is a slow growing benign swelling in lateral neck and hence can be confused with other benign swellings of neck. We present a case of ancient schwanoma of cervical sympathetic chain which masqueraded as carotid body tumour highlighting the importance of contrast enhanced CT and MRI pre-operatively along with restricted utility of FNAC in correct diagnosis of lateral neck swelling.

Keywords: Ancient schwannoma, Cervical sympathetic chain, Carotid body tumour

## **INTRODUCTION**

Schwannomas are benign tumours arising from the nerve sheath cell. 25-40% of schwannomas are found in head and neck. The most common site is parapharyngeal space from lower cranial nerves and very rarely from cervical sympathetic chain. These tumours are slow growing encapsulated tumours which mimic benign neck swellings and should be differentiated pre-operatively to avoid surgical surprises. We present case of 50 year old female with ancient schwannoma of cervical sympathetic chain which masqueraded as carotid body tumour. This case highlights the drawback of FNAC and indespesibilty of MRI in correct diagnosis of neck swellings.

#### **CASE REPORT**

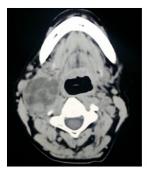
50 year old female presented with a 4 cm firm, globular gradually progressive swelling in the right side of the neck since 2 years. The swelling was prominent just anterior to anterior border of sternocleidomastoid but was

going beneath it. It was smooth surfaced, regular in margins, non-pulsatile and non-tender. There was presence of transmitted pulsation with no audible bruit. The mass had horizontal mobility but no movement in the vertical plane suggestive of the origin from a linear vertically oriented structure. There was no neurological deficit or abnormality.



Figure 1: Globular lateral neck swelling present inferior to angle of mandible and arising deep to sternocleidomatoid muscle and extending beyond the anterior border of sternocleidomatoid.

CT scan was suggestive of a soft tissue swelling with diffuse contrast enhancement with areas of degeneration.



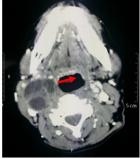


Figure 2: (a) CT neck showing a globular soft tissue swelling with areas of degenerations. (b) On contrast, the swelling does not enhance intensely but the areas of degeneration and separations become prominent.

The carotid artery is pushed anteriorly.

MRI was showing a T1 isointense with enhancement on administration of contrast and T2 hyperintense mass which is suggestive of schwannoma. FNAC was not diagnostic of any pathology.

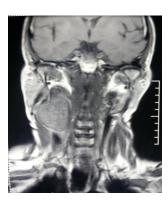
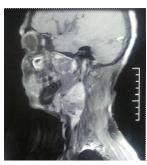


Figure 3: T1 weighted MRI (coronal plane) depicts a hypointense soft tissue density in lateral neck with clear demarcation of fat plane on the lateral and medial side.



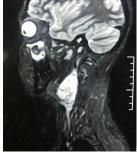


Figure 4: (a) T1 weighted post contrast image (sagittal plane) shows diffuse uptake of contrast which washes off slowly with areas of low uptake (b) T2 weighted MRI images (sagittal plane) shows a heterogenous but hyperintense soft tissue swelling suggestive of neural origin.

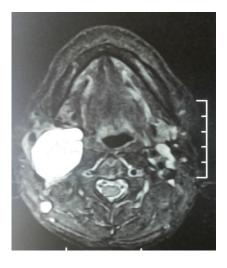


Figure 5: T2 weighted MRI (axial cuts) demonstrates a hyperintense smooth and regular soft tissue swelling along with a tail (red arrow) from the source of origin, most probably a nerve.

On surgical exploration, the mass was delineated and was seen arising from cervical sympathetic chain pushing the great vessels anteriorly. Vagus and phrenic nerves were identified and preserved. Complete tumour removal was done with sacrifice of cervical sympathetic chain. Patient developed post-operative Horner syndrome.

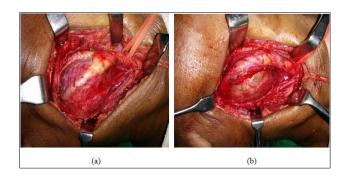


Figure 6: Carotids (blue arrow) are pushed anteriorly by the mass with vagus (green arrow) being shifted to a more antero-lateral position.



Figure 7: (a) The specimen of the schwannoma after surgical removal with the stump of the cervical sympathetic chain. (b) The cut section of the schwannoma which shows multiple areas of degeneration with septations.

Histopathology report revealed the characterstics suggestive of schwannoma but with features of degeneration especially loss of Antoni A areas. A presence of atypical cells with large, polymorphic, bizarre and hyperchromatic nuclei but absence of mitotic figure suggestive of ancient schwannoma.

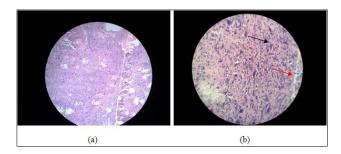


Figure 8: The histopatholgy shows hypercelluarity with irregular nuclei (red arrow) but no evidence of mitosis. There are areas of hyalinization (black arrow) with hyperchromatism suggestive of degenerative changes. These are the typical feature of ancient schwannoma. On immune-histochemistry, the tumour was intensely positive for S 100.

#### **DISCUSSION**

Schwannomas are benign encapsulated tumors which arise from schwann cells. These tumours were first described by Verocay in 1910. 25-40% of all schwannomas are found in head and neck area. These tumors are slow growing tumors which may manifest as a painless, smooth regular mass. The retrostyloid compartment of the parapharyngeal space is the most common site of origin.<sup>1</sup>

Schwannomas arising from cervical sympathetic chain are very rare. Schwannomas are generally solid tumour but may have increased hyalinization and degenerated areas due to progression in size over a long duration and are termed as Ancient Schwannomas. Ackerman and Taylor<sup>2</sup> suggested the terminology schwannoma to connote the long duration and degenerative changes to the appearance of these lesions. . It is characterized by nuclear hyperchromasia, mild nuclear pleomorphism, stromal edema, fibrosis, and xanthomatous changes leading to a misdiagnosis of malignancy in the aspirates. Secondary degenerative changes due to the long duration resulting in waxing and waning of the tumor size.<sup>3</sup> Degenerative changes such as hemorrhage, calcification, and fibrosis are commonly seen in schwannoma, but cystic changes are rare. Degeneration is due to central tumor necrosis as the schwannoma grows to a size beyond the capacity of its blood supply. The tumor as such may be infiltrated with large number of siderophages. Such tumors are generally seen in retroperitoneum<sup>3</sup> and relatively rare in neck.

Ancient schwannomas are confused with many cystic masses of the neck due to its degenerated areas with non-

diagnostic hemorrhagic aspirate. The most common lesions which are the differential diagnosis in this area of neck are tubercular lymphadenopathy, carotid body tumour (as was in our case), lymphangioma or soft tissue sarcoma. The best tool to diagnose such neck masses where FNAC may falter more often than not is MRI scan with administration of gadolinium contrast.

In our case, due to presence of increased vascularity on CT and MRI of neck with presence of hemorrhagic aspirate, the patient was referred to us as carotid body tumour from peripheral hospital.

But when we studied the MRI scan, it showed that the internal and external carotid has been pushed anteriorly and the internal jugular vein was pushed slightly anterolaterally. There was not significant splaying of carotids. This is the main difference between schwannomas arising from cervical sympathetic chain and carotid body tumours. <sup>4,5</sup> Both carotid body tumours and schwannomas of vagus nerve splay the carotids and don't push them anteriorly. DWI (diffusion weighted imaging) is great help in differentiating a schwannoma from carotid body tumour as schwannomas will have a delayed wash-out of contrast relative to carotid body tumour.

In the literature, the CT appearance of a schwannoma has been described as a well circumscribed, inhomogeneous mass of low density, which can be explained by the following microscopic pattern: hypocellular areas (Antoni type B) adjacent to more cellular regions (Antoni type A) and cystic degeneration.<sup>5</sup>

Surgical excision remains the mainstay of treating this benign condition although there is a lack of evidence to support this in view of the rarity of this disease entity.<sup>7</sup>

# **CONCLUSION**

Ancient schwannoma of cervical sympathetic chain is very rare entity and can present as carotid body tumour due to the site of location but can be differentiated by displacement of vessels seen on contrast enhanced CT scan where carotid body tumour will pre-dominantly splay the vessel but schwannoma of cervical sympathetic chain will displace the vessel anteriorly than splaying significantly. MRI is of tremendous help in such swelling as carotid body tumours will have significant flow voids but schwannoma wont and hence, is an indispensible diagnostic tool. Role of FNAC is not very significant in such swellings.

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