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## Case report

### ANCIENT SCHWANNOMA OF THE CERVICAL VAGUS NERVE: A RARE BENIGN NEUROGENIC TUMOR

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#### ABSTRACT

Schwannomas are benign, slow-growing encapsulated tumors that originate from Schwann cells of the nerve sheaths. Schwannoma originating from the cervical vagus nerve is an extremely rare neoplasm that usually occurs in men between the 3rd and 6th decades of life. Ancient schwannoma is a rare variant that was first described by Ackerman and Taylor in 1951 as a long standing degenerative schwannoma that presented with wide areas of hyalinized matrix. A case of ancient Schwannoma of the cervical vagus nerve in a 56-year-old male who presented to our department with history of a firm and painless mass lesion on right side of the neck is being reported here.

**Keywords:** Ancient schwannoma, Vagus nerve, Computed tomography, Head and neck.

#### INTRODUCTION

Schwannomas are neurogenic tumors arising from schwann cells of neural sheath. Schwannomas are the most common types of neurogenic tumors found in the head and neck. Common sites for schwannomas in the neck are the vagus nerve, less commonly the glossopharyngeal nerve, the ventral and dorsal cervical nerve roots, the cervical sympathetic chain, and the brachial plexus<sup>1</sup>. Radiological Imaging plays a significant role when evaluating a parapharyngeal space mass<sup>2</sup>. Ancient schwannoma is a uncommon variant that was first described by Ackerman and Taylor in 1951<sup>3</sup>, which is an encapsulated tumor with benign nature. It is characterized by degeneration and diffuses hypocellular areas. These changes are believed to occur because it takes a long time for schwannomas to develop<sup>4</sup>. It is characterized by degenerative changes typified by perivascular hyalinization, calcification, cystic necrosis, relative loss of Antoni type A tissue, and degenerative nuclei

that may be misinterpreted as sarcomatous pleomorphisms<sup>5</sup>. However, the absence of mitosis and the presence of cystic necrosis and a well-defined capsule without surrounding invasion helps to differentiate these lesions from high-grade lesions<sup>6</sup>. The cause of schwannomas is not clear, although it appears genetics play a role. Most schwannomas are sporadic, but about 10% are associated with familial neurofibromatosis type 2. The goal of treatment is complete excision with possible preservation of the involved nerve<sup>7</sup>.

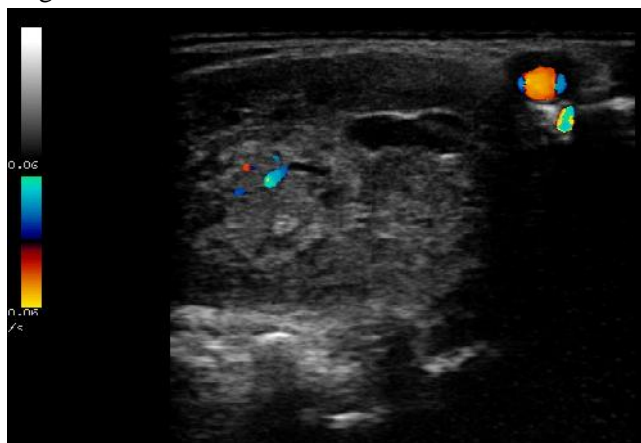
#### CASE REPORT

A 56 Yr old male patient presented with history of painless swelling in right side of neck since 3 years. Department of Radiodiagnosis, Mysore Medical College and Research Institute, Mysore.

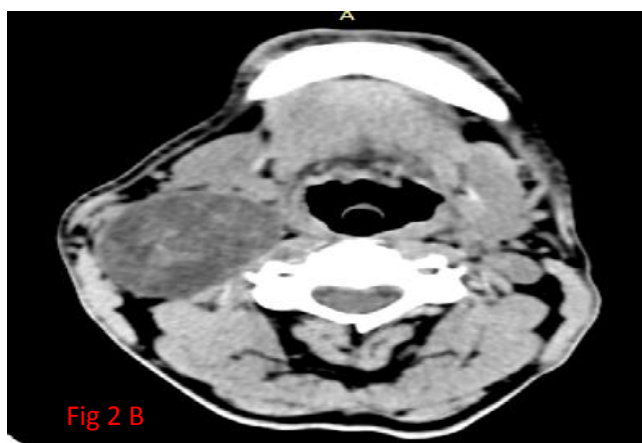
Color Doppler ultrasound of the neck [figure1] revealed a 6cm x 4.5cm x 3.3cm heterogeneous mass with no evidence of internal vascularity. The internal Jugular vein was compressed and the carotid artery was significantly displaced by the mass lesion.

Computed tomography scan [figures 2-5] revealed a well defined heterogeneously enhancing mass lesion with few non enhancing cystic areas within, on right side of the neck extending from the level of the C2 vertebra to the level of the D2 vertebra, measuring 6.5cm x 5.5cm x 4.2cm, displacing the carotid artery anteriorly and compressing the right internal jugular vein.

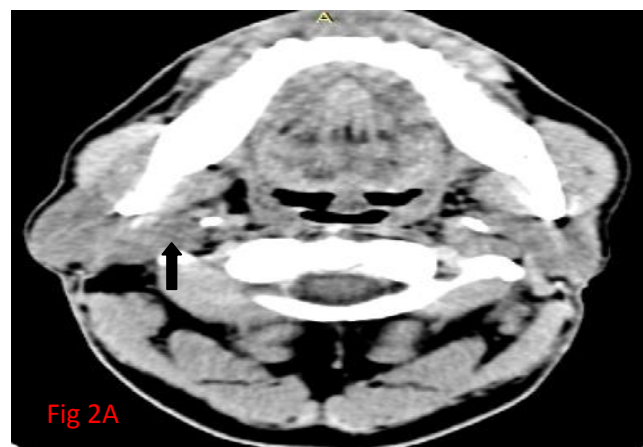
Histopathology showed spindle cells in clusters. These cells showed moderate degree of nuclear pleomorphism with bland chromatin. Fibrillary intercellular stroma was noted. No necrosis or mitosis was seen. Correlating with the long standing nature of the lesion and radiological evidence of well defined borders favoured a benign neck mass, and hence a diagnosis of ancient schwannoma was made.



**Fig 1:** Ultrasonography of the neck on right side showing an heterogeneous mass lesion displacing and compressing internal jugular vein and common carotid artery.

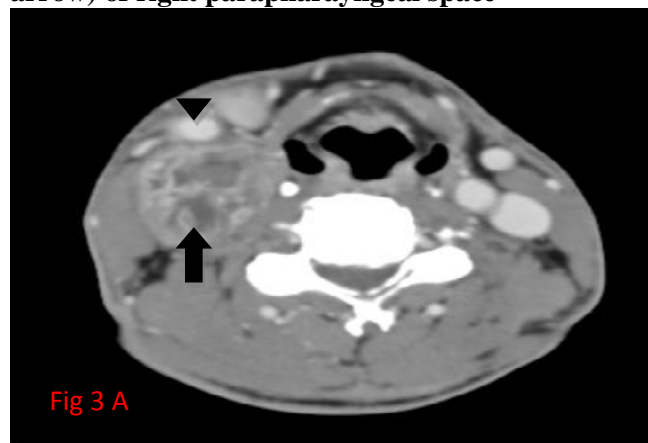


**Fig 2 B**

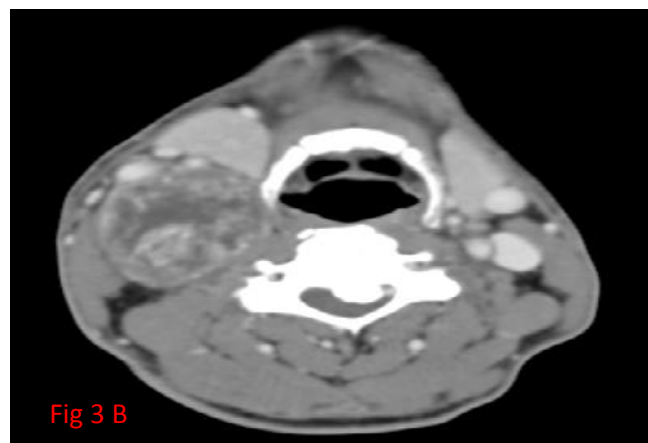


**Fig 2A**

**Figure 2: (A, B) Axial unenhanced CT scan shows a heterogeneous, predominantly hypodense mass lesion in the post styloid compartment (black arrow) of right parapharyngeal space**

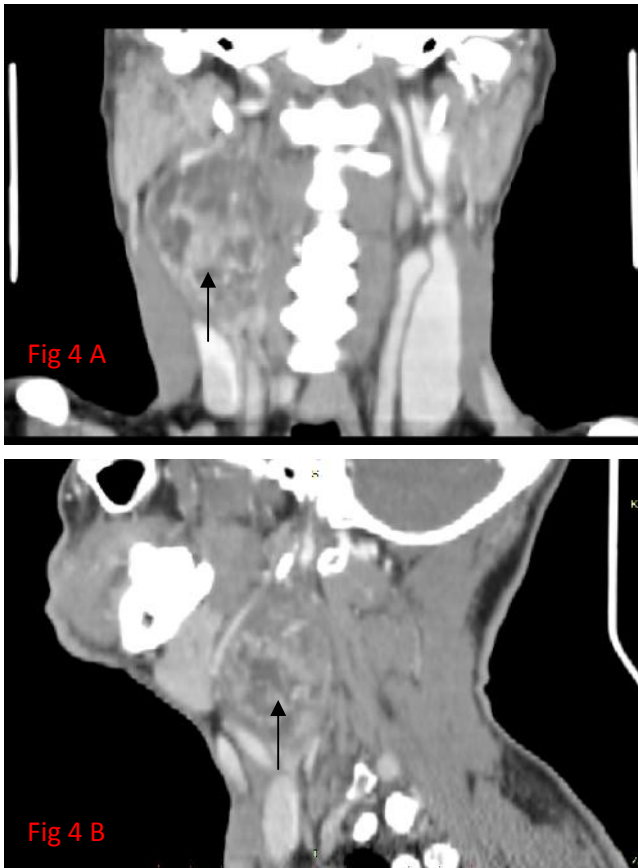


**Fig 3 A**



**Fig 3 B**

**Figure 3 (A, B) Axial contrast enhanced images show a heterogeneously enhancing mass lesion (arrow) with few non enhancing areas displacing the right internal carotid artery anteriorly (black arrowhead).**



**Fig 4: (a) Coronal reformatted image, (b) sagittal reformatted image shows a heterogeneously enhancing mass lesion in right side of the neck(arrow).**

## DISCUSSION

A schwannoma is a benign nerve sheath tumor composed of schwann cells, which normally produce the insulating myelin sheath covering the peripheral nerves. In the parapharyngeal space of neck, schwannomas may arise from the last four cranial nerves or the autonomic nerves, the vagus being the more common site. The differential diagnosis of a parapharyngeal space mass is based on the division of the space into prestyloid and post-styloid compartments. The prestyloid compartment contains the parotid gland, fatty tissue, and lymph nodes. The post-styloid compartment contains the carotid sheath with the sympathetic chain and cranial nerves IX through XII. Thus, masses arising in the post-styloid compartment include carotid artery lesions, paragangliomas arising from the vagus nerve or the carotid body, neurogenic tumors involving cranial nerves IX to XII, or sympathetic chain neurogenic lesions<sup>1</sup>. Most schwannomas are fairly homogeneous soft tissue masses and appear hypodense or isodense

to skeletal muscle on noncontrast CT and tend to be hypointense or isointense to skeletal muscle on T1-weighted MR images and variably hyperintense on T2-weighted images. Despite their hypovascularity, they enhance significantly on both CT and MR images and can mimic a paraganglioma. The enhancement of the schwannomas is seen at least 2 minutes after the contrast injection and depicts the equilibrium phase of the contrast agent and the poor venous drainage of the tumor. Dynamic scans can reveal the true nature of the lesion and differentiate it from hypervascular lesions. The enhancement pattern of neural tumors can vary; it may be an inhomogeneous enhancement (owing to necrosis and hemorrhage) or even lack of enhancement<sup>1</sup>.

When evaluating a parapharyngeal space mass which helps in guiding surgical approach for the mass lesion, the most important landmarks to note in terms of imaging are (a) the deep portion of the parotid gland and the stylomandibular tunnel region; (b) the ICA, its size, shape, and the direction of any displacement; (c) the direction of any displacement of the fat of the prestyloid compartment; (d) the effect of a mass on the surrounding structures, including the pharynx, masticator space, mandible, and skull base; and (e) the size of the mass<sup>2</sup>.

Ancient schwannoma, a degenerative neurilemmoma, a rare variant of schwannoma first described by Ackerman and Taylor in 1951<sup>3</sup>, is a schwannoma subtype characterized by degeneration and diffuse hypocellular areas. These changes are believed to occur because it takes a long time for schwannomas to develop<sup>4</sup>. The radiologic findings for ancient schwannoma are similar to those for schwannoma, which on contrast-enhanced computed tomography shows enhancement in capsules or pericystic areas<sup>5</sup>. It is characterized by degenerative changes typified by perivascular hyalinization, calcification, cystic necrosis, relative loss of Antoni type A tissue, and degenerative nuclei that may be misinterpreted as sarcomatous pleomorphisms. However, the absence of mitosis and the presence of cystic necrosis and a well-defined capsule without surrounding invasion helps to differentiate these lesions from high-grade lesions<sup>5,6</sup>.

Ancient schwannomas are benign tumors and the goal of treatment is complete excision with possible preservation of the involved nerve as complete resection is usually curative with a good prognosis<sup>7</sup>.

## CONCLUSION

Ancient schwannomas are rare benign tumors, radiological and histological findings in correlation with long standing nature of the lesion will aid in the pre operative diagnosis and proper management. The goal of treatment is complete excision with possible preservation of the involved nerve.

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**Conflict of Interest:** Nil

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