



# Multiple Ancient Schwannoma of the Accessory Nerve: A Case Report

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## Abstract

We are reporting a case of multiple ancient schwannoma of the accessory nerve in a 57 year old male. Accessory schwannomas are rare with only 19 cases reported in literature so far. Ancient schwannoma is even rarer in the head and neck region with no other cases associated

with the accessory nerve being reported in literature so far.

## Keywords

*schwannoma, neurilemoma, ancient, accessory nerve, neck*

## Introduction

Schwannoma is an infrequent, benign, encapsulated tumour arising from nerve sheath cells<sup>(1)</sup>. They are generally smooth and uniform in gross appearance and are usually surrounded by or attached to their nerve of origin. Although more commonly encountered in the extremities, approximately 25% of the cases originate from the neural structures of the head and neck region. Histologically five variants of schwannomas have been described: common, plexiform, cellular, epithelioid, and ancient schwannoma<sup>(2)</sup>.

The term ancient schwannoma was first suggested by Ackerman and Taylor<sup>(3)</sup> in a review of 48 neurogenic tumours of the thorax. They reported 10 cases which showed similar features of a typical schwannoma, but distinctive because significant portions of these tumours were composed of only few cells within a hyalinized matrix. They clarified that these features occurred in schwannoma of long duration, and hence coined the term 'ancient schwannoma'. This type is characterized by diffuse areas of hypocellularity, focal accumulations of hyaline material, and fatty degeneration<sup>(4)</sup>. In addition to thick capsules and relative absence of necrosis, they are usually infiltrated by large numbers of siderophages and histiocytes. The histopathology findings favouring a diagnosis of ancient schwannoma were neoplasms composed

of cellular areas made up of ovoid to spindle cells, having eosinophilic cytoplasm and hyperchromatic nuclei with nuclear pallasading, cystic degeneration, sheets of foamy and hemosiderin laden macrophages and areas of hemorrhage. Similar histopathologic features were present in both the masses.

Since that first report, a few authors described ancient schwannoma in a variety of locations in the head and neck region ranging from the oral cavity, parotid, submandibular salivary glands and even the orbit. They had their origins from the cranial nerves and the parapharyngeal space.

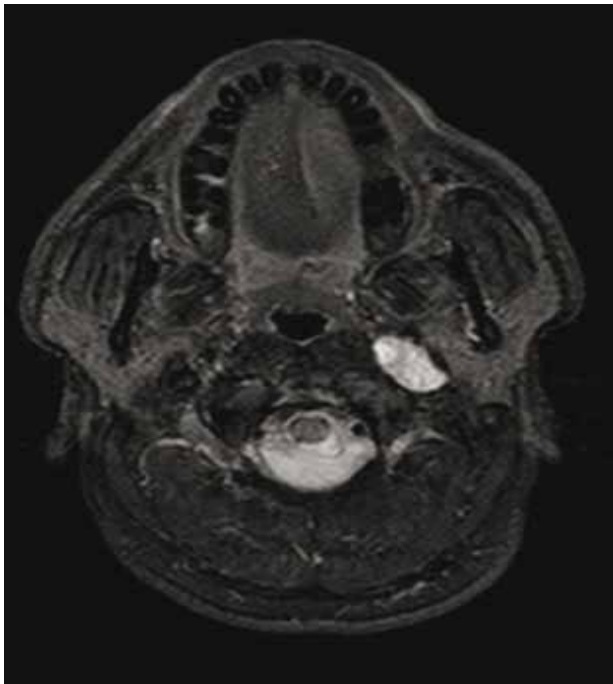
## Case History

A 57-year old gentleman was referred to our out patient clinic with history of swelling in the upper part of left side neck of six months duration with rapid increase in size during the last one month. He also complained of upper neck pain originating posterior to the ear and radiating down to the lower neck and along the armpit and the upper limb along with numbness and parasthesia. On examination there was a 3x3 cm smooth swelling in the upper part of neck on the left side deep to sternomastoid. It was mobile and tender. An ill defined swelling approximately measuring 4x3 cm in the left supra scapular region overlying the trapezius was also noticed. Atrophy of the sternomastoid and trapezius muscles on the ipsilateral side, with difficulty in shoulder abduction, along with atrophy of the tongue was noticed on clinical

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examination. The gag reflex on the same side was weak and on indirect laryngoscopy both vocal cords were found to be mobile.

**Imaging:** An MRI scan which showed an 8x6 cm elongated lobulated mass lesion extending from right jugular foramen to C4 vertebrae along the jugular vein. It showed solid and cystic components which expanded the jugular foramen but with out any intra cranial component. Ipsilateral internal jugular vein was compressed and internal carotid artery was displaced anteriorly without encasement of the artery. Hypoglossal canal was normal. Following contrast administration the mass showed moderate enhancement of the solid component. Left sigmoid sinus and internal jugular vein were not visualized probably due to compression. It was reported as left lower cranial nerve schwannoma. There was a diffuse mass in the supraclavicular area measuring 4x3 cm.

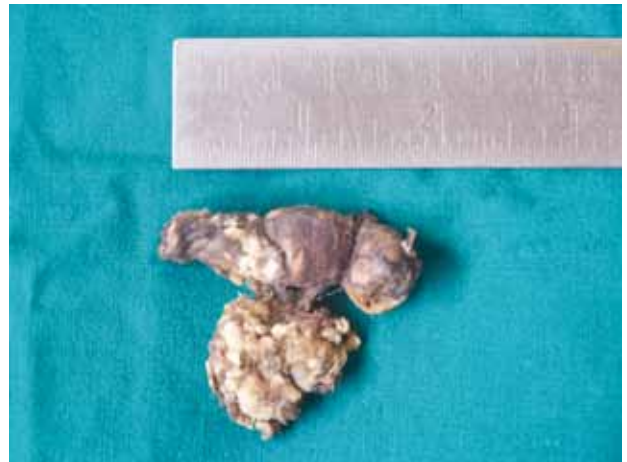


**Fig. 1 :** Axial view showing the mass

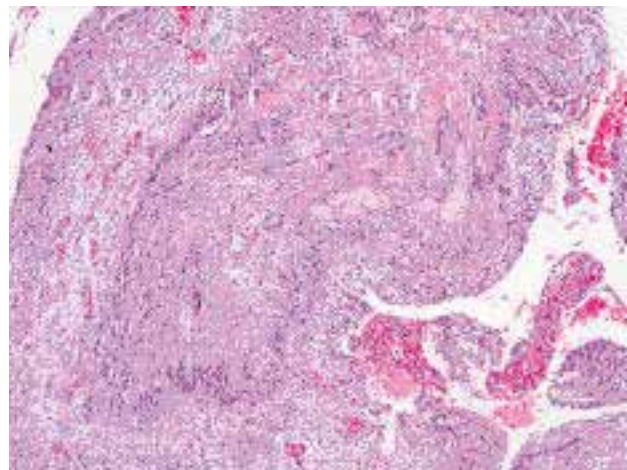
The neck mass was excised through an upper apron incision and on exploration there was a soft yellowish lobulated mass arising from the accessory nerve causing thickening of the nerve from the level of entry into sternomastoid muscle to the jugular foramen. There was another soft, cystic swelling of size 4cm, within the trapezius muscle in the suprascapular region, which was also excised.

## Pathology

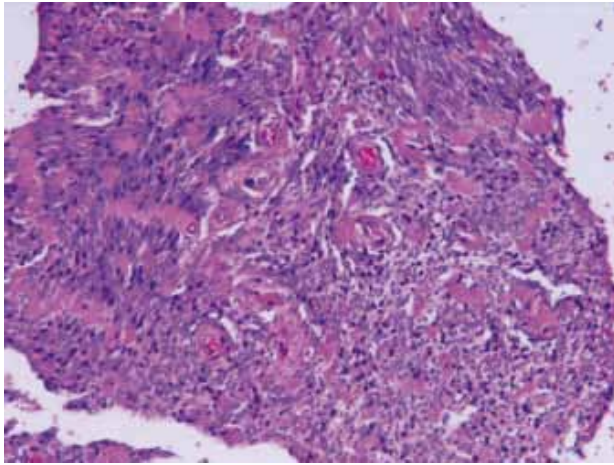
The mass from the upper neck measured 5x2.5x1 cm, cut section showed grayish yellow solid area with multiple small cystic spaces, the second mass overlying the trapezius measured 4x2.5x1.5 cm, cut section was similar to the first mass. Microscopic examination of both the masses showed a neoplasm composed cellular (Antoni A) areas made up of ovoid to spindle cells having eosinophilic cytoplasm and hyper chromatic nuclei with nuclear pallsading and less cellular Antoni B areas. Cystic degeneration, sheets of foamy and heamosiderin laden macrophages and areas of hemorrhage were noted. With these features the diagnosis of ancient schwannoma was made.



**Fig. 2a :** Gross specimen showing nodular lobulated soft tissue mass measuring 5x2.5x1 cm with greyish yellow solid areas with multiple cystic spaces.



**Fig. 2b :** Microphotograph showing cellular area of the tumor with nuclear pallsading (Antony A) area surrounded by sheets of foamy macrophages, capillaries, areas of hemorrhage & cystic change (H&E 100)



**Fig. 2c : Higher magnification of the cellular area showing nuclear palisading. (H&E X 200)**

### **Discussion**

We are presenting this case for its rarity. This is the first reported case of multiple ancient schwannoma of the accessory nerve. Schwannoma and neurofibroma are the two most frequently encountered forms of nerve sheath tumours. Over the years a variety of names have been used to differentiate between nerve sheath tumours; these include neurofibroma, schwannoma, neuroma, neurilemmoma and neurinoma. The current view is that schwannomas arise from the progenitor of the Schwann cell, whereas neurofibromas arise from a mesenchymal origin closer to a fibroblast<sup>5</sup>. Schwannomas are often solitary and are composed solely of Schwann cells of peripheral sensory nerves, whereas neurofibromas are often multiple as seen in Von Recklinghausen's neurofibromatosis<sup>6</sup>. The peak incidence for nerve sheath tumours is between 20 and 40 years of age and it occurs equally in both sexes, they can occur anywhere within the spine, the thoracic spine being the most frequently affected. However, lesions in the cervical area present at a much younger age compared to lesions of the thorax. The reason for this is not yet obvious, although one hypothesis is that the upper cervical roots are much larger compared to other nerve roots and any tendency to abnormality might be expected in the area of highest nerve cell density, and therefore might appear at a younger age<sup>5</sup>.

MR Imaging is the imaging of choice in the assessment of the peripheral nerve sheath

tumours. It has proved to be equal or superior to Computed Tomography in the evaluation of soft tissue tumours<sup>7</sup>. It involves no ionizing radiation and has multiplanar capability. It allows the assessment of the nerve sheath tumour because the longitudinal growth of the tumour along the axis of the nerve can be shown using the coronal or sagittal planes. There is overlap between the radiological features of benign and malignant nerve sheath tumour<sup>8</sup>. The size of the lesion is not an indication of malignancy. MR imaging signal heterogeneity, contrast enhancement, peripheral edema and adjacent muscle atrophy are the differentiating features of malignancy

Approximately 25–30% of all schwannomas occur in the head and neck area<sup>9</sup>, whilst ancient schwannoma rarely affects the head and neck region. In their original description, Ackerman and Taylor<sup>3</sup> proposed that the ancient schwannomas begin as a diffuse cellular overgrowth with increased vascularization, followed by decreased vascularity with resulting hyalinization. Especially in frozen sections, the areas with hypercellularity and atypia may mislead the pathologist to a diagnosis of a malignant lesion. Prior to the realization that the nuclear atypia and hyperchromatism observed in ancient schwannomas were not a sign of malignancy but rather a regressive phenomenon, many of these lesions were erroneously diagnosed as sarcomas<sup>10</sup>.

Head and neck schwannomas are sometimes misdiagnosed and preoperative investigations are not often fruitful<sup>11</sup>. Although aspirates of ancient schwannoma may show some features such as nuclear pleomorphism, nuclear inclusions, perivascular sclerosis, xanthomatous changes, or nuclear atypia<sup>10</sup>, fine-needle aspiration biopsy has questionable value<sup>12</sup>. It may show unclear histopathological results from these cases which have the potential to confuse this lesion with a sarcoma. In this case, although the FNAC diagnosis was of a schwannoma, the final diagnosis of an ancient schwannoma could be made only at the final histopathological examination.

## Conclusion

A case of an ancient schwannoma is described. This type of tumour is uncommon in the head and

neck region and extremely rare in the accessory nerve. Fine needle aspiration biopsy may be misleading. Complete excision leads to cure.

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