



A rare case of *schwannoma* at carotid space of neck region mimicking as *paraganglioma*.

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Abstract-

This is a case of carotid body tumour at right side of neck region with initially diagnosed as *paraganglioma*. A 22 year old female patient with an history of pain and swelling at right side of neck region with intermittent headache difficulty during deglutination. Patient has taken medication for pulmonary kochs for 8 months. Now presented with the mentioned complaints with a swelling at right side swelling at neck region. The features and the correlation with MRI report diagnosed as *paraganglioma* and then on *Histopathologic* examination showed a tumour mass composed of two different areas admixed with each other either alternatively or diffusely. The final pathological impression was *Neurilemmoma* (*Schwannoma*). The diagnosis was definitive with no evidence of *paraganglioma*.

Keywords: *paraganglioma*, *Schwannomas*, *benign tumors*, *Arbud*

Introduction-

Schwannomas are benign tumors of nerve sheath origin that most often arise from the superior vestibular branch of the eighth cranial nerve in the cerebellopontine

angle.^[1] However, *schwannomas* may rarely originate from the ninth or tenth cranial nerves within the jugular foramen^[2]. Besides *schwannomas*, *paragangliomas* may arise from scattered collections of paraganglial tissue, and *meningiomas* may form from an *intraforaminal* connective tissue layer. Additional considerations include metastatic tumor, primitive *neuroectodermal tumor*, and jugular vein pseudomas.

The imaging characteristics of these lesions are often similar. However, *paragangliomas* are classically and clinically diagnosed, associated with on magnetic resonance imaging (MRI) because of multiple areas of signal void interspersed with hyperintense foci due to narrowed flow voids. Accordingly, *paragangliomas* are highly vascular on angiography^[4]. In contrast, *schwannomas* are typically described as avascular or hypovascular on angiography, without central flow voids on MRI^{[2],[3],[5]}. We describe an unusual case of schwannoma at right carotid space which seems as a paraganglioma as earlier, A schwannoma with prominent central flow voids on MRI and present an overview of the relevant imaging findings.

Case report-

A 22 year old female patient with an complaints of pain and swelling at right side of neck region with intermittent headache and difficulty during deglutination. Patient has taken medication for pulmonary kochs for 8 months. Now presented with the mentioned complaints with a swelling at right side swelling at neck region, right side hearing loss with tinnitus and dizziness.

A contrast-computed tomography (CT) scan of her head was performed for further evaluation and revealed there was 2.4 x 3.1 x 3.5 cm sized well defined enhanced mass lesion in right carotid space causing splaying of internal and external carotid arteries however right (internal carotid artery) and (external carotid artery) show normal contrast opacification without any luminal narrowing. Few central non-enhancing areas noted within. This lesion appears to be supplied by external carotid artery likely right ascending pharyngeal artery. Anteriorly, abutting right submandibular gland and external carotid artery. Laterally, displacing right internal jugular vein with maintained fat plane. Medially abutting the greater cornu of the hyoid bone on right side without its erosion or sclerosis. Superiorly, reaching upto the level of right transverse process of C2 vertebral body. Inferiorly abutting the bifurcation of the right common carotid artery causing splaying of internal and external carotid arteries. On impression, there was well defined enhancing mass lesion in the right carotid space with suggestive of neoplastic etiology likely neurogenic in origin.

Subsequent MRI examination showed a 2.5 x 3.2 x 3.8 cm (AP x ML x SI) sized well defined homogenously enhancing soft tissue mass lesion in right carotid space causing splaying of common carotid artery bifurcation with external, internal carotid arteries coursing along peripheral aspect. It extends from C2-C4 vertebral levels superoinferiorly. Anteriorly, it was

abutting the submandibular gland with maintained fat planes. External and internal carotid arteries are coursing along anterior and posterior aspect of the lesion, however show normal flow voids. External carotid artery appears mildly narrowed. Posteriorly, it compressing the internal jugular vein, however it showed normal flow voids. Laterally, it abutting the sternocleidomastoid muscle and parotid gland with maintained fat planes. Medially it reaching upto the parapharyngeal space, however there was no pharyngeal luminal compression. A small approximately 2.2 x 2.0 x 1.1 cm sized lesion showing similar morphology appears to extend from lesion anteriorly and extending posterolateral to the Internal Jugular vein. On impression, small well defined homogenously enhancing soft tissue mass lesion in right carotid space causing splaying of common carotid artery bifurcation with morphology and extensions. Features are more likely suggestive of paraganglioma or nerve sheath tumour like schwannoma.

An elective operative approach was selected. Excision of carotid body tumour performed under general anaesthesia. A large Carotid tumour of approximately 3 x 3.5 x 3 cm removed. Common carotid artery identified. Carotid pulses clearly seen. Internal and external carotid arteries clearly seen. Carotid body tumour was situated at the carotid bifurcation. Superior lateral planes of the tumour well identified and origin of the carotid arteries are well identified. Lymphatic structures of the tumour separated. Internal and external carotid arteries are dissected away from the tumour. Then the posterior aspect of the tumour dissected. Vagus nerve and the hypoglossal nerve preserved. Haemostasis achieved. Replacement of the structures done. The tumour was situated almost at the posterior aspect of carotid artery. Delicately removal of carotid body tumour performed and the layerwise closure done.

The patient recovered appropriately postoperatively with proper medications and dressing, and was discharged on postoperative day 7. There was good improvement from her preoperative clinical symptomatology.

Histopathologic examination showed a tumour mass composed of two different areas admixed with each other either alternatively or diffusely. The *hypercellular* areas are comprised of spindle shaped cells with spindle to small vesicular nuclei; arranged in palisading pattern and also revealing verocay body formations in scattered areas. Another areas are of *hypocellular* nature and show oedematous foci with ill defined cystic spaces and with no evidence of malignancy. The final pathological impression was *Neurilemmoma* (Schwannoma). The diagnosis was definitive with no evidence of *paraganglioma*.

Discussion-

Paragangliomas, also called glomus jugular tumors when located in the Jugular Foramina, comprise the majority of primary Jugular Foramina tumors^{[3][6]}. Paragangliomas are said to follow the paths of least resistance, affecting the mastoid air cells, vascular channels, auditory tube, and neural foramina^[5]. They may also proliferate into the tympanum, causing destruction of the ossicles and/or the bony labyrinth^[3]. The characteristic spread of the tumor leads to a “moth-eaten” pattern of destruction of the temporal bone, which is evident on CT-bone windows. Medial extension and extracranial extension into the carotid space is less common^[3].

Paragangliomas are highly vascular tumors which show strong heterogeneous enhancement with contrast agents^[4]. Mentioned vascularity is manifested with prominent flow voids, resulting in the classic “salt-and-pepper” appearance. Angiography may reveal a coarse tumor

blush with derivation from the ascending pharyngeal artery and the presence of early venous drainage^[3].

Jugular Foramina schwannomas, in contrast to paragangliomas, result in scalloped, sclerotic expansion rather than lytic destruction of the temporal bone^[2]. Four presentations of Jugular Foramina schwannomas were identified by Samii et al^[7]: type A: primary cerebellopontine angle involvement, type B: primary JF involvement, type C: primary extracranial involvement, and type D: both intracranial and extracranial components resulting in a dumbbell-shaped lesion. Middle ear involvement is rare^[2].

On CT imaging, mass lesion in right carotid space causing splaying of internal and external carotid arteries however right (internal carotid artery) and (external carotid artery) show normal contrast opacification without any luminal narrowing.

On MRI, mass lesion in right carotid space causing splaying of common carotid artery bifurcation with external, internal carotid arteries coursing along peripheral aspect. It extends from C2-C4 vertebral levels superoinferiorly. Anteriorly, it was abutting the submandibular gland with maintained fat planes. External and internal carotid arteries are coursing along anterior and posterior aspect of the lesion, however show normal flow voids. External carotid artery appears mildly narrowed. Posteriorly, it compressing the internal jugular vein, however it showed normal flow voids. Laterally, it abutting the sternocleidomastoid muscle and parotid gland with maintained fat planes. Medially it reaching upto the parapharyngeal space, however there was no pharyngeal luminal compression.

They do not generally exhibit central flow voids, distinguishing them from paragangliomas, and are correspondingly avascular or hypovascular on angiography^[2].

Our case represents an atypical presentation of carotid space schwannoma. Initially, paraganglioma was suspected because of normal or narrowed flow voids on MRI, as well as angiographically evident tumor blush. Additional features that also supported paraganglioma the epidemiologic preponderance of paragangliomas in the JF or near to it.

However, on reevaluation of neuroimaging, evidence supporting schwannoma could also be identified, particularly in the pattern of growth. There was a scalloped instead of a “moth-eaten” lytic pattern of bony involvement with limited expansion into the mastoid air cells in addition, there was extensive medial extension into the hypoglossal canal and clivus as well as extracranial extension into the carotid space (Samii type D), features less commonly seen in JF paragangliomas^[3]. Moreover, the enhancement pattern on CT and tumor blush on angiography was less prominent that would be expected for a paraganglioma, although the vascularity of the tumor ultimately dissuaded us from a diagnosis of schwannoma.

Schwannomas with prominent vascularity have been previously described in the literature, first in the 1970s via angiography^{[8], [9]}. More recent reports have described their appearance on MRI, though they only rarely resemble the findings presented in this report. Yamakami et al^[10] presented 5 cases of hypervascular vestibular schwannomas with the primary finding of large peripheral flow voids representing large draining veins; whereas 2 of 5 cases showed intratumoral flow voids, their appearance lacked the distinct pattern of hypointensity and hyperintensity characteristic of a “salt & pepper” appearance.

In fact, a ‘salt & pepper’ appearance of intratumoral flow voids in schwannomas, though the cases presented were located

extracranially^[11]. Though previous reports attributed the peripheral flow voids of some schwannomas to enlarged draining veins, Kato ascribes the central vascularity described in his report to the degenerative change seen in “ancient schwannomas,” such as abnormal calcification and hyalinization.

However, our case was devoid of these changes on imaging and histopathologic examination, perhaps indicating that a tumour mass composed of two different areas admixed with each other either alternatively or diffusely. The hypercellular areas are comprised of spindle shaped cells with spindle to small vesicular nuclei; arranged in palisading pattern and also revealing verocay body formations in scattered areas. Another areas are of hypocellular nature and show oedematous foci with ill defined cystic spaces and with no evidence of malignancy. The final pathological impression labelled as Neurilemmoma (Schwannoma).

In conclusion, our case represents the only report of a carotid space schwannoma with normal or narrowed flow voids on MRI. Though this appearance has been traditionally ascribed to paragangliomas, schwannoma should also be considered in carotid space lesions. Ultimately, the pattern of tumor expansion may provide important insights in the imaging diagnosis.

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