Peripheral Nerve Sheath Tumor of Cervical Vagus Nerve

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By N.U. Bahri, MD [1], Ketan Rathod, MD [2], and Dimple Shah, MD [3]

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Case history: A 45-year-old female patient was referred to our hospital with painless swelling on left side of neck. She had been aware of the swelling for the last eight months which was gradually increasing in size. Initial examination revealed an evident swelling with smooth contour on left side of neck. No sign of inflammation of overlying skin was noted. On palpation, it was non-pulsatile, soft to firm in consistency and was not adherent to overlying skin. The swelling was not tender. Clinically it was suspected to be an enlarged lymph node. Cytological examination was non-conclusive. The patient was then referred to radiology department for CT scan.

Non contrast CT images demonstrated an oblong lesion in left carotid space with well-defined boundaries and a homogenous low density (average 16 HU) (Figure 1).
**Figure 1:** Non-contrast axial CT image: fluid attenuation lesion in left carotid space (blue arrow). On post contrast CT, the lesion did not show any significant enhancement (Figure 2).

**Figure 2:** Contrast axial CT image: Non enhancing lesion in left carotid space (blue arrow). The lesion appears to displace common carotid and its external and internal branches medially and internal jugular vein laterally (Figure 3).
Figure 3: Contrast axial CT image: non-enhancing lesion causing separation of common carotid artery and internal jugular vein (red arrows).

Because of the location, characteristic displacement of cervical major vessels, provisional diagnosis of peripheral nerve sheath tumor of cervical vagus nerve was considered. However, because of the non-enhancing nature of the lesion, possibility of 2nd branchial cleft cyst was also considered as a differential diagnosis. So, ultrasound examination with colour doppler study was performed for further evaluation.

On ultrasound, the lesion showed solid appearing peripheral hypoechogenicity and central hyperechogenicity (Figure 4).
Figure 4: Ultrasound: Solid heterogeneously hypoechoic lesion on left side of neck. Vagus nerve was not clearly delineated.

On color Doppler study, presence of internal vascularity was noted within (Figure 5). The lesion showed displacement of carotid arteries medially and internal jugular vein laterally.
Figure 5: Color Doppler imaging: Significant internal vascularity was noted. To further characterise this lesion, MRI was done. T2WI and STIR images showed homogenous hyperintensity (Figure 6 & 7).
Figure 6: Axial T2WI: Hyperintense lesion in left carotid space (blue arrow).
**Figure 7:** Coronal Short Tau Inversion Recovery Image: Homogenously hyperintense lesion (blue arrow)
T1WI showed homogenous hypointensity (Figure 8).
Figure 8: Axial T1WI: Homogenously hypointense lesion (blue arrow)
So, further contrast MRI was performed. On contrast studies, the lesion showed heterogenous and progressive delayed enhancement (Figure 9 and 10).
Figure 9: Contrast axial image: (early phase); Early mild heterogenous enhancement is noted. (blue arrow)
Figure 10: Contrast axial image: (delayed); Progressive delayed enhancement is noted. (blue arrow)
Wide local excision was done under general anaesthesia. The lesion was well encapsulated and was easily separated from vagus nerve (nerve of origin). No post-operative Horner’s syndrome was noted. Gross specimen appeared well-circumscribed, firm rubbery mass.

Intraoperative photograph of the lesion following excision from the parent nerve.
Figure 13: Haematoxylin and eosin stained section shows neural and fibrous elements intermingled in loose stroma. Findings were suggestive of neurofibroma.

**Diagnosis:** Peripheral Nerve Sheath Tumor of Cervical Vagus Nerve

In our case, the lesion simulated Type II Branchial cyst on plain and contrast enhanced CT scan and non-contrast MRI study. However, ultrasound played an important role which clearly depicted it as a solid lesion with internal vascularity. Contrast MRI study further confirmed the diagnosis of peripheral nerve sheath tumor of cervical part of vagus nerve.

**Discussion:** Neurogenic tumors can be divided into three groups according to their tissue of origin:[3]

- Peripheral nerve: (schwannoma, neurofibroma, malignant schwannoma)
- Sympathetic ganglia: (ganglioneuroma, ganglioneuroblastoma, neuroblastoma)
- Paraganglia: (chemodectoma, pheochromocytoma)

25 – 45% of nerve sheath tumors occur in head and neck region. In the neck region, these tumors may arise from vagus nerve, sympathetic chain or cervical nerve roots including the brachial plexus. [1]

The vagus nerve exits the posterior cranial fossa through the jugular foramen; before entering the thorax, it runs its entire course in the neck within the carotid sheath. Tumors arising from this cervical segment of the vagus nerve are rare and are often accidentally detected at surgery, with most being asymptomatic [5,6]

Neurofibromas arise either from nerves of dermis, subcutaneous tissue or other deeper tissues. They are most commonly seen in patients between the ages of 20 and 40 years. Most neurofibromas are slowly growing painless masses causing no particular symptoms until they reach a considerable size. The nerve of origin is usually incorporated within the neurofibroma and a fusiform configuration is typically seen on gross inspection. Cervical neurofibromas may be solitary lesions (although rare) or they may be seen in patients with Neurofibromatosis1 (Von Recklinghausen’s disease) [4]
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Ultrasound features: both schwannomas and neurofibromas show a heterogenous hypoechoic pattern. A pseudocystic appearance with posterior enhancement is commonly seen with both tumors and this may lead to confusion. Neurofibromas however, do not always show posterior enhancement. Neurofibromas may be lobulated and schwannomas may show true well defined cystic component within the tumor. The contours of the tumor are well defined; irregular borders may suggest malignant changes. The nerve immediately adjacent to the tumor may be thickened producing tapering appearance to the tumor. Presence of nerve thickening is the best clue to the nature of these tumors. Histologically, schwannomas are more vascular than neurofibromas. Colour doppler imaging also demonstrates this increased vascularity, which is extremely sensitive to pressure. Even mild transducer pressure obliterates intra-tumoral vascularity. Despite the histological differences between neurofibromas and schwannomas, sonographic features have not allowed reliable distinction between the two.\[9\]

The typical appearance of peripheral nerve sheath tumor on MRI appears hyperintense on T2WI; the lesion can appear heterogenous due to haemorrhagic or cystic degeneration. After the administration of contrast agent marked contrast enhancement may lead to a misdiagnosis of hypervascular tumor. [2] Actually, peripheral nerve sheath tumor is a relatively hypovascular lesion and the enhancement is due to extravascular leakage through abnormally permeable vessels with poor venous drainage.

Typical signs on MRI:
- The “split-fat” sign - displacement of fat in the neurovascular bundle by the slow-growing tumor, and seen as a thin rim of hyperintense signal on T1 weighted sequences.[10]
- The “fascicular” sign - nerve fascicles can be seen as multiple ring like structures within.[10]
- The “target” sign - low signal intensity with a ring of high signal intensity peripherally on T2WI.

This pattern reflects the histologic features of the tumor with peripheral myxomatous tissue with surrounding a fibrocollagenous core.[10]

Imaging findings of schwannomas are similar to those seen with neurofibromas and, in many cases, cannot be distinguished. However, some features can help differentiate these two lesions. When the parent nerve is identified, an eccentrically positioned lesion (in relation to the nerve) suggests a schwannomas, whereas a centrally located mass suggests a neurofibroma. Heterogeneous appearance with degeneration and cystic cavitation are much more common in schwannomas than in neurofibromas. “Ancient” schwannomas refer to long-standing lesions with advanced degeneration exhibiting calcification, hyalinization, and cystic cavitation, findings that can be identified on imaging.[11]

Metastatic lymph nodes, paragangliomas, and schwannoma of the cervical sympathetic chain are the usual differential diagnoses to be considered. Metastatic lymph nodes are often multiple with a known primary facilitating the diagnosis [6] Paragangliomas can be differentiated by dynamic contrast study: a schwannoma will always show slow steady increase enhancement while paragangliomas show rapid enhancement.[2] Another important point of differentiation is displacement of neck vessels: splaying of the carotid bifurcation with hypervascularity suggests a carotid body tumor, whereas in cases without hypervascularity, a cervical sympathetic chain schwannoma is another possibility. Vagus nerve schwannomas can separate the internal jugular vein and internal carotid artery, but seldom widen the carotid bifurcation. [7,8]

The accepted treatment of a cervical nerve sheath tumor is surgical excision, and less often an observational approach is adopted. The benefits of surgical excision must be carefully weighed against the risk of postoperative neurologic deficit especially in view of the indolent course seen with many of these tumors. Hence, an accurate and prompt diagnosis of cervical nerve sheath tumors is crucial. [6]

On gross examination, neurofibromas are white gray and lack degenerative changes. Less frequently, these tumors may arise in and expand a large nerve to form fusiform mass, when if confined by the epineurium, a true capsule may be present. There is prominent increase in endoneurial myxomatous material that separates myelinated and non-myelinated axons; a disorderly proliferation of fusiform Schwann cells, fibroblasts and axons as well as perineurial fibrous thickening. [12]

In summary, there are certain imaging characteristics that may aid the radiologist in establishing a focused preoperative diagnosis of a peripheral nerve sheath tumor and differentiating from other solid lesions of carotid space.

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