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Radiodiagnosis

RADIOLOGICAL FINDINGS OF TUMOURS ARISING FROM NERVE SHEATH AND AUTONOMIC NERVOUS SYSTEM

Dr. Sathiyamoorthy Jothy* MBBS, MD (RD), Faculty, Department of Radio-Diagnosis,Government Theni Medical College & Hospital, Theni- 625512 *Corresponding Author

Dr Muthuraj Subramanian MBBS, MS, Faculty, Department of Surgery,Government Theni Medical College & Hospital, Theni- 625512

ABSTRACT Tumours arising from the peripheral nerve sheath tumours are categorized into benign and malignant tumours. Benign nerve sheath tumours are neurofibroma and schwannoma. Peripheral nerve sheath tumours account for nearly 12% of benign and 7%-8% of malignant soft tissue neoplasms. Ultrasound, CT and MRI are imaging modalities that are useful in diagnosis of peripheral nerve sheath tumours. Choice of modality varies with location of lesion. In this case series, we describe five rare cases of which three are nerve sheath tumors in different locations and two are tumours arising from autonomic nervous system.

KEYWORDS : Magnetic Resonance Imaging, Neurofibroma, Schwannoma , Paraganglioma, Neuroblastoma

INTRODUCTION:

Tumours arising from the peripheral nerve sheath tumours are categorized into benign and malignant tumours. Benign nerve sheath tumours are neurofibroma and schwannoma. Peripheral nerve sheath tumours account for nearly 12% of benign and 7%-8% of malignant soft tissue neoplasms.

Radiological study is important to assess the anatomic origin and to find out the size and extent of the tumour and for differential diagnosis.

DISCUSSION

Tumours arising from nerve sheath are neurofibroma, Schwannoma and malignant nerve sheath tumours. Tumours arising from ganglia of autonomic nervous system include paraganglioma (which arise from neuroendocrine cells) and neuronal tumours (neuroblastoma, ganglioneuroblastoma and ganglioneuroma)[1,2].

Schwannoma

Schwannomas are encapsulated neoplasms that originate in nerve sheaths and are usually slow growing. Chest wall schwannomas arise from spinal nerve roots and intercostal nerves . It occurs in the age between 20 and 50 years. Small tumors tend to be spheroid, firm, and well circumscribed , whereas larger tumors are ovoid or irregularly lobulated. In radiographs bone erosion or scalloping can occasionally be seen. In non-enhanced CT scans, Schwannoma typically shows a well-circumscribed homogeneous mass with attenuation slightly less than or equal to that of muscle. On CT scans acquired after contrast material administration, the attenuation of the mass is equal to or slightly greater than that of muscle. The signal intensity of schwannoma on T1-weighted MR images is equal to or slightly greater than that of muscle and on T2-weighted images is markedly greater, with increased contrast between the high-signal-intensity nerve sheath tumor, intermediate-signal-intensity fat, and low-signal-intensity muscle. The nerve from which the tumor originated can often be seen along one side of the mass.The presence of bone erosion without destruction indicates the benign nature and slow growth rate of this lesion.

Neurofibroma

Neurofibromas are slow-growing neoplasms that originate from a nerve, may or may not be encapsulated, and may include components of cystic degeneration and calcification. Neurofibromas develop most commonly in patients between the ages of 20 and 30 years, in men and women equally. Radiographs of the spine may show a widening of neural foramina because of tumor extension along spinal nerve roots. Most neurofibromas are hypoattenuated on nonenhanced CT scans and show heterogeneous enhancement after intravenous administration of contrast material. Many neurofibromas have a histologic pattern of zonal distinction, with a central zone composed of a highly cellular component and a peripheral zone composed of abundant stromal material, which results in a target like appearance on T2-weighted MR images. On these images, the mass is characterized by a rim of increased signal intensity that surrounds the central part of the tumor, which has a lower signal intensity .

Paraganglioma

Paraganglioma may arise from sympathetic ganglia including adrenal medulla (pheochromocytoma) or in sympathetic chain (paraganglioma)[1,2]. Paraganglioma may also arise from parasympathetic chain along 9th, 10th cranial nerves, examples being glomus jugulare and carotid body tumour. Mediastinal paragangliomas occur 1) in the middle mediastinum, near origins of aorta and pulmonary artery or 2) in the posterior mediastinum (paravertebral paraganglioma).[2] On plain CT, they are typically heterogeneous and show intense enhancement after administration of intravenous contrast.

TABLE-1: RADIOLOGICAL FINDINGS

TUMOUR	RADIOLOGICAL FINDINGS
neurofibroma, schwannoma	Rib erosion, well defined contour, extra skeletal, location
paraganglioma , ganglioneuroma	Location in the paravertebral region Intense enhancement in paraganglioma.

CASE 1

Neurofibroma/ Schwannoma

A 61 years old man presented with cough and chest pain. His CT of chest shows an extrapulmonary lesion in right lower hemithorax. It is homogeneously hypodense with smooth margin. Sagittal and coronal images show its extrapleural location in relation to anteroinferior aspect of the rib above implying that it runs in intercostal groove. It measures about 3.0 x 1.5 cm. Follow-up CT after 6 months shows stable size. The imaging features are suggestive of benign nerve sheath tumour arising from intercostal nerve.[3]

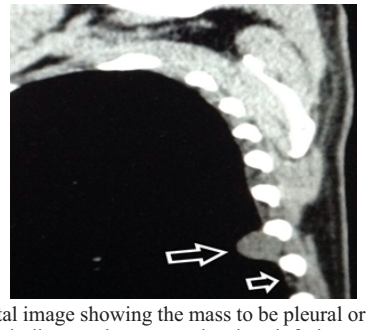

Fig 1 : Sagittal image showing the mass to be pleural or extrapleural. Long arrow indicates the mass abutting inferior aspect of rib; intercostal neurovascular bundle is not seen separately. Short arrow points normal intercostal neurovascular bundle as dots in the next intercostal space.

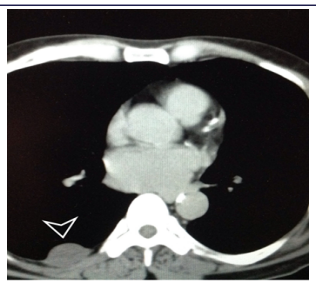

Figure 1a: Another axial image showing smooth margin of the lesion which is homogeneous.

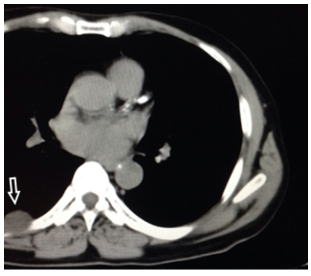

Fig 1 b: showing hypodense lesion with smooth margin in posterior aspect of thorax in right side, close to rib.

Case 2: paraganglioma

CT chest of a 50 years old man shows a homogenous hyperdense mediastinal mass in middle mediastinum. It shows intense contrast enhancement.It is located beneath aortic arch in the region of aortico-pulmonary window. Imaging features are suggestive of mediastinal paraganglioma.[4] Other possibility is hemangioma.

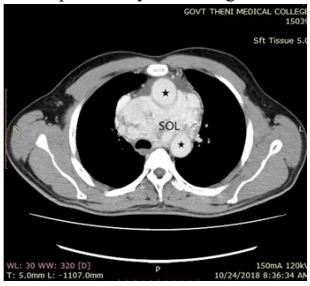

Figure 2a: Axial CT image of thorax with intravenous contrast shows intensely enhancing mass (marked as SOL) in middle mediastinum beneath aortic arch. Star mark indicates ascending and descending aorta.

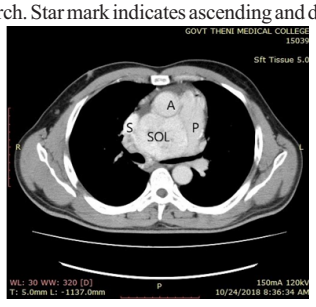

Figure 2b: Another axial image showing relationship of the mass (SOL) to SVC (S), Aorta (A) and main pulmonary artery (P).

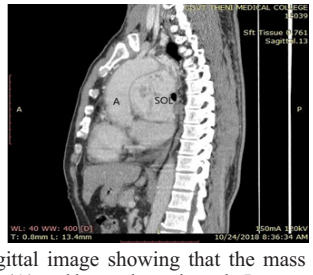

Figure 2c: Sagittal image showing that the mass (SOL) is behind ascending aorta (A) and beneath aortic arch. Intense enhancement and location at aorto-pulmonary window are highly suggestive of paraganglioma

Case 3: neuroblastoma

An one year old girl baby was brought with abdominal distension. On examination the baby was hypertensive. CT Abdomen shows: retroperitoneal soft tissue density nodular lesions with calcifications. Visualized sections of Thorax shows posterior mediastinal soft tissue density lesion with calcifications and anterior mediastinal soft tissue density with calcification. Features are consistent with neuroblastoma.[5]

Figure 3a: CT abdomen (without intravenous contrast): axial image shows retroperitoneal soft tissue lesions with calcification.

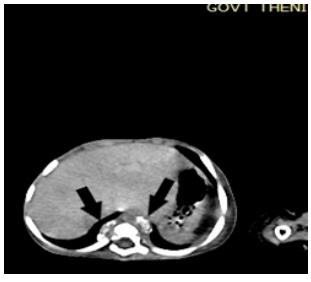

Figure 3b: Axial CT sections through lower thorax shows posterior mediastinal soft tissue density lesion with calcifications

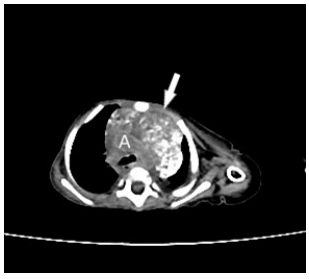

Figure 3c: Axial CT sections through upper thorax show anterior mediastinal soft tissue density lesion with calcifications seen anterior to aortic arch.

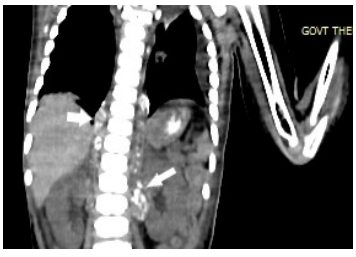

Figure 3d: CT image: Coronal section showing posterior mediastinal (upper arrow) and retroperitoneal (lower arrow) soft tissue lesion with calcification. Lesions are distributed along the course of sympathetic ganglia suggesting diagnosis of neuroblastoma.

Case 4: Schwannoma.

A 35 years old lady presented with swelling in right side of neck below angle of mandible for the past 6 months. It was painless and there was no fever. Cervical lymphadenopathy was clinically suspected and patient was referred for ultrasound of neck. Ultrasound shows a well-defined homogenous hypoechoic solid mass with vascularity. It is located lateral to right common carotid artery bifurcation and is lateral to internal carotid artery without splaying carotid bifurcation. It is posterior to internal jugular vein. The lesion is contiguous with right vagus nerve with thickening of vagus nerve. Prominent vessels are seen in and around vagus nerve. Imaging features are suggestive of Vagal Schwannoma.[6]

Figure 4a

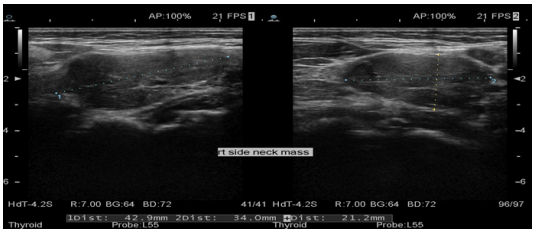

Figure 4a: Ultrasound of neck showing ovoid isoechoic lesion with smooth margin in lateral aspect of upper part of neck mimicking enlarged lymph node.

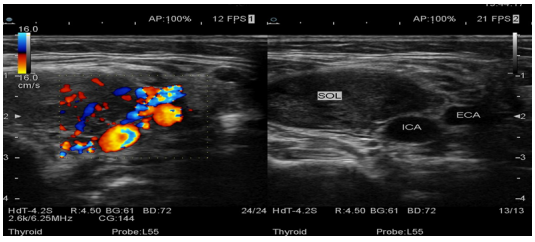

Figure 4b: Ultrasound with colour Doppler shows that the mass is lateral to internal carotid artery and external carotid artery. That the mass does not splay internal & external carotid artery differentiates vagal tumors from carotid body paraganglioma.[7] Vascularity is seen in the mass. Internal jugular vein is displaced anteriorly. That the mass is lateral to carotid artery distinguishes vagal Schwannoma from Schwannoma of cervical sympathetic chain.[8,9]

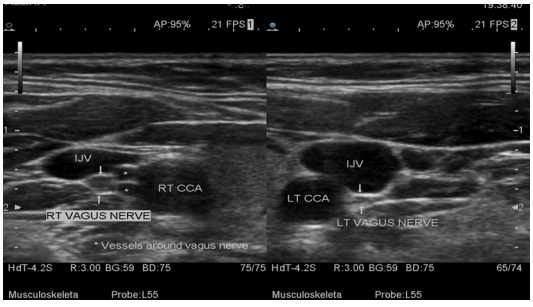

Figure 4c: Ultrasound done with high frequency probe (5 to 13 MHz linear probe) showing thickened right vagus nerve (marked by arrow marks). Prominent vessels (*) are noted around vagus nerve (between the nerve and common carotid artery). Abbreviations: CCA- Common carotid artery IJV- Internal jugular vein.

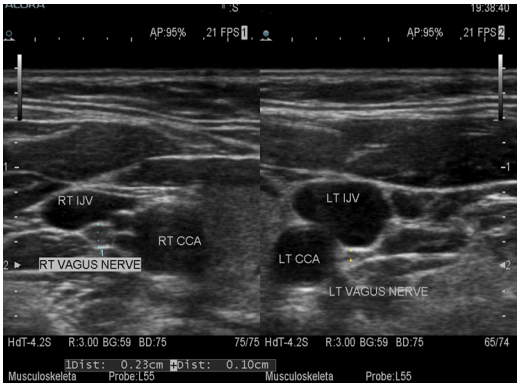

Figure 4d: Thickening of right vagus nerve shown objectively (comparison made with left side normal vagus nerve)

Case 5: Neurofibroma/ Schwannoma

A 30 years old lady with swelling in left side of neck in parotid region whose ultrasound shows elongated hypoechoic lesion superficial to left parotid gland; it is homogeneously hypoechoic with posterior acoustic enhancement. Some internal vascularity reveals that it is a solid lesion. It is about 4 cm long and 1 cm in cross sectional diameter. There is a tail-like elongation of the lesion caudally. A small part of cranial part of the lesion dips beneath parotid capsule into parotid gland. Imaging features are suggestive of nerve sheath tumor (arising from a branch of facial nerve). Operative findings were concordant revealing benign nerve sheath tumour.

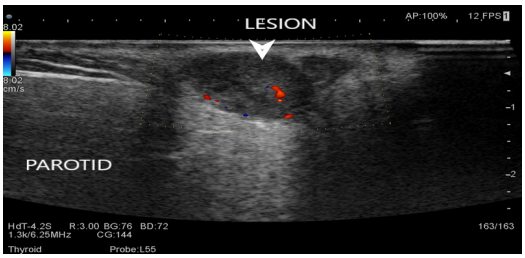

Figure 5a: Ultrasound image shows well-defined hypoechoic lesion superficial to left parotid gland with posterior acoustic enhancement. Internal vascularity implies that it is a solid lesion.

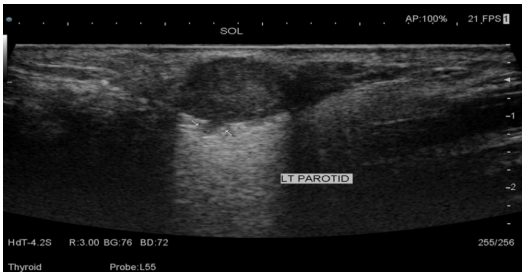

Figure 5b: Ultrasound done with high frequency (5-13 MHz) linear probe showing that a small part of cranial part of the lesion dips beneath parotid capsule into parotid gland.

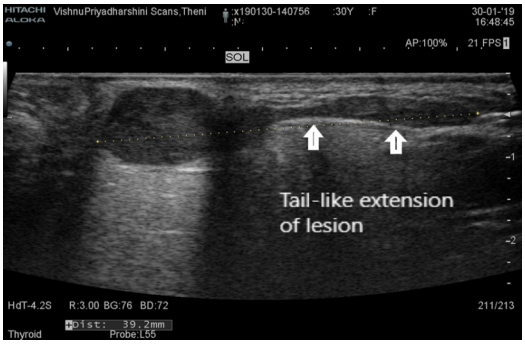

Figure 5c showing tail-like extension of the lesion suggesting nerve sheath tumour.[10]

CONCLUSION:

From this case series, the following teaching points are emphasized:

1. Possibility of nerve sheath tumour is to be included in the differential diagnosis of extrapulmonary lesions near pleura. In addition to axial CT images, sagittal and coronal images also should be studied. Smooth margin, relation to intercostal space (especially neurovascular bundle) are some of features that need to be assessed.
2. When intensely enhancing lesion is seen in mediastinum or in relation to expected location of ganglia (in paravertebral region, or around aorta/ pulmonary artery) is found, possibility of paraganglioma is to be considered.
3. When we see posterior mediastinal lesions, neurogenic tumours (either nerve sheath tumor or neuronal tumor) must be considered among the differentials. When we see posterior mediastinal lesion in a child, either neuroblastoma or ganglioneuroma must be included in the differential diagnosis.
4. In evaluation of neck mass, careful analysis of relation of lesion to great vessels and adding colour Doppler screening with Ultrasound or contrast enhanced CT/ MRI of neck are useful in differentiating tumours arising from autonomic ganglia (paraganglioma) in carotid sheath (Carotid body glomus tumour and glomus vagale) and tumours arising from nerves (nerve sheath tumors) from more common cervical lymphadenopathy.
5. Facial nerve neurofibroma is one among the differentials for a swelling in preauricular/parotid region (apart from parotid tumor and lymphadenopathy). Ultrasound finding of tail-like extension of the lesion is useful clue for diagnosing nerve sheath tumour.

REFERENCES:

1. Vinay Kumar, Abul K. Abbas, Nelson Fausto, (2004). 7th edition, Robbins and Cotran PATHOLOGIC BASIS OF DISEASE, Philadelphia: Elsevier; 2004. Page: 789 and pages: 1218,1219
2. H. Page McAdams, Jeremy J.Erasmus at al (2003). 'Mediatinum' in John R. Haaga, et al, 4th edition, CT and MR Imaging of the Whole Body. Missouri: Mosby. Volume 1 Chapter: 29
3. Tateishi, U., Gladish, G.W., Kusumoto, M. et al, Chest wall tumors: radiologic findings and pathologic correlation: part 1. Benign tumors. Radiographics. 2003;23:1477–1490
4. John D. Pavlus , Brett W. Carter , Marc D. Tolley , Elaine S. Keung , Leila Khorashadi , and John P. Lichtenberger 3rd . Imaging of Thoracic Neurogenic Tumors. American Journal of Roentgenology. 2016;207: 552-561. 10.2214/AJR.16.16018
5. Papaioannou G., McHugh K. Neuroblastoma in childhood: review and radiological findings. Cancer Imaging. 2005;5:116–127
6. Anil T. Ahuja, Yolanda Y.P.Lee & H.Y. Yuen, Infrahyoid Carotid Space Vagal Schwannoma, Infrahyoid Carotid Space, (2014). Anil T. Ahuja. Diagnostic ultrasound: Head and Neck. Philadelphia: Elsevier. Chapter: 79. II 5: 32 -35
7. A.B. Rao, K.K. Koeller, C.F. Adair, From the archives of the AFIP. Paragangliomas of the head and neck: radiologic-pathologic correlation Armed Forces Institute of Pathology. Radiographics, 19 (November–December (6)) (1999), pp. 1605-1632 Review. PMID: 10555678
8. D.M. Saito, C.M. Glastonbury, I.H. El-Sayed, D.W. Eisele, Parapharyngeal space Schwannomas: preoperative imaging determination of the nerve of origin, Arch Otolaryngol Head Neck Surg., 133 (July (7)) (2007), pp. 662-667 PMID: 17638778
9. M. Furukawa, M.K. Furukawa, K. Katoh, M. Tsukuda, Differentiation between Schwannoma of the vagus nerve and Schwannoma of the cervical sympathetic chain by imaging diagnosis Laryngoscope, 106 (December (12 Pt 1)) (1996), pp. 1548-1552 PMID: 8948621
10. Soldatos, Theodoros & Fisher, Stephen & Karri, Sirisha & Abdulrahman, Ramzi & Sharma, Rohit & Chhabra, Avneesh. (2015). Advanced MR Imaging of Peripheral Nerve Sheath Tumors Including Diffusion Imaging. Seminars in musculoskeletal radiology. 19. 179-190. 10.1055/s-0035-1546823.