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Retroperitoneal Neoplastic Lesion (A Rare Case of Extra Adrenal Paraganglioma)

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Abstract: ***The purpose was to highlight the diagnosis and treatment of extra-adrenal para-gangliomas, which often causes catecholamine hypersecretion and hypertension. Extra-adrenal retroperitoneal paragangliomas (PGLs) arise from dispersed paraganglia that tend to be symmetrically distributed in close relation to the aorta and sympathetic nervous system.***

Keywords: Para ganglioma, extra adrenal, schwannoma, retroperitoneum, Retroperitoneal tumour

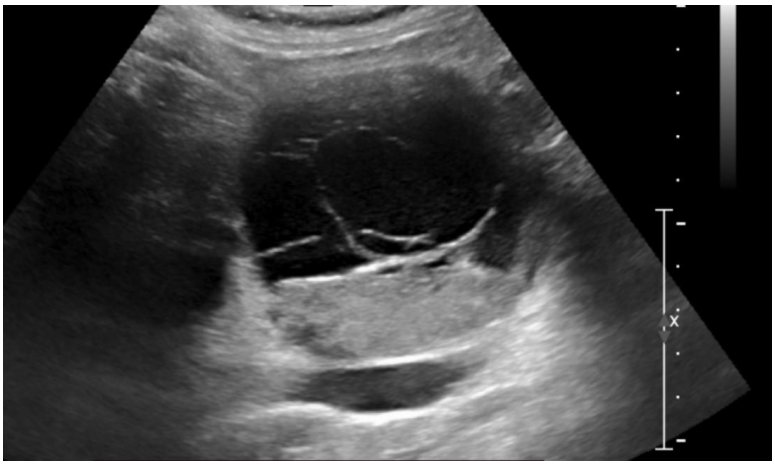
1. Aim and Objective

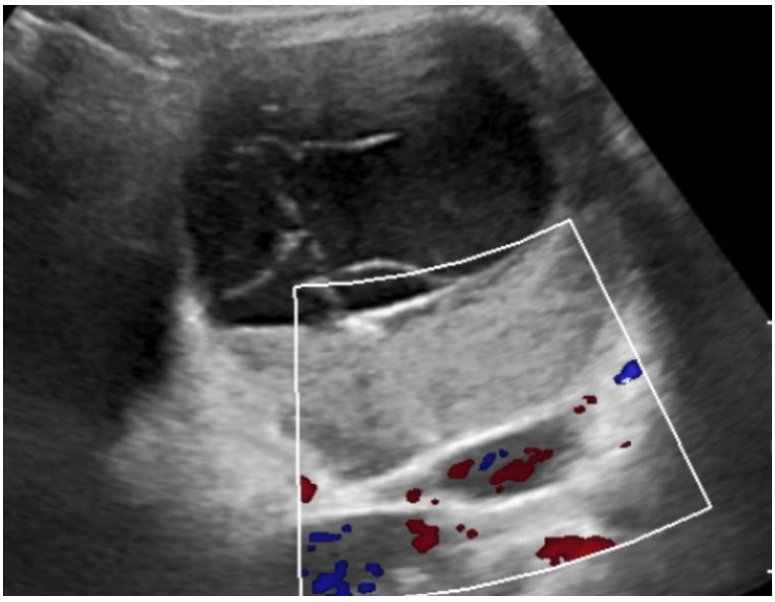
We aim to describe a case report of an incidental finding of left retroperitoneal paraganglioma in a young female who presented with right flank pain.

1) To determine the organ of origin.
2) To define the extent and involvement of adjacent structure.
3) To determine the diagnostic accuracy of CT scan.

2. Imaging Findings

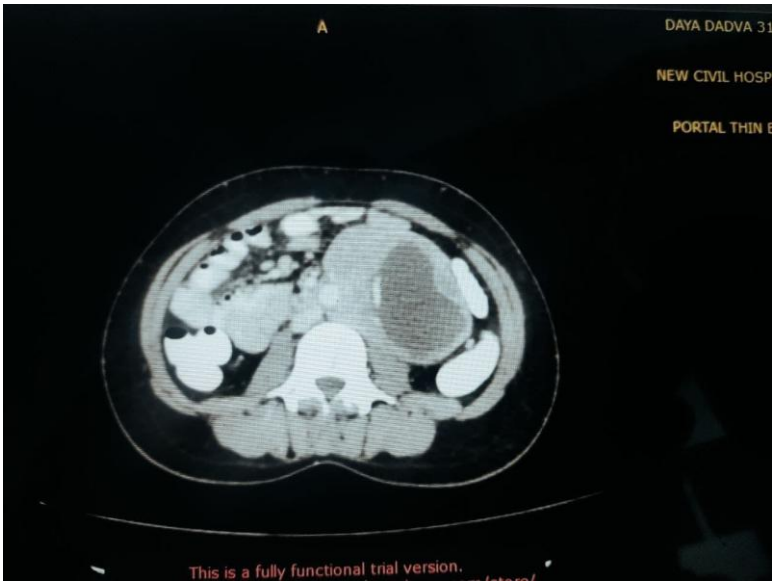
on USG, there is lobulated solid cystic lesion, with evidence of few septations and internal vascularity in solid component noted adjacent to inferior pole of left kidney and aorta, involving / compressing upper ureter resultant moderate hydronephrosis on left side.

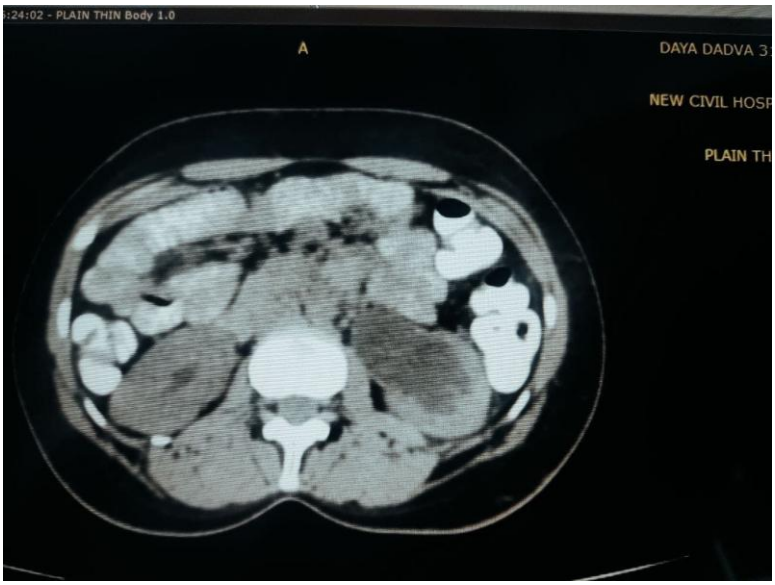




On CECT abdomen, Approx. (81 x76 x66) (AP X MLX SI) mm sized Lobulated soft tissue density lesion with subtle calcification with central (69 x43) mm sized cystic component noted in left lumber region at L3-L4 vertebral level, extending from inferior pole of left kidney up to aortic bifurcation which show heterogenous postcontrast enhancement with feeding arterial channels.

Above mentioned lesion incases inferior mesenteric artery and closely abuts abdominal aorta medially, with area of contact 170-180 degree, however no evidence of definitive invasion noted.





It also abuts left psoas muscle and prevertebral space inferiorly with preserved fat palne, closely abuts jejunal loops with indistinct fat plane anteromedially.

Above mentioned lesion involve proximal 1/ 3, upper ureter resultant moderate hydronephrosis of left kidney, however left kidney shows normal post contrast enhancement.

Excretion of contrast noted in left pelvicalyceal system in 10 minutes delay scan however rest of ureter not visualized on subsequent delay scan. (Poorly excreating kidney).

On histopathology, the sections show well encapsulated mass composed of varying sized nests, anastomosing cords, trabecular and organoid arrangement of neoplastic cells separated by highly vascularized fibrous septa. Individual cells show mild to moderate pleomorphism having round/oval nuclei, finely granular (salt and paper chromatin) with moderate to abundant amount of eosinophilic cytoplasm. Typical and atypical mitosis figures (>3/10 hpf) with areas of hemorrhage are seen. Vascular invasion and capsular invasion are seen. Necrosis is not seen. All the margins are involved by tumor. Overall histological and immunohistochemistry findings suggestive of Extra adrenal Paraganglioma.

3. Discussion

a) Background: Extra-adrenal retroperitoneal paragangliomas (PGLs) arise from dispersed paraganglia that tend to be symmetrically distributed in close relation to the

aorta and sympathetic nervous system. They are rarely encountered in every day surgical practice. Meanwhile, Extra-adrenal retroperitoneal paragangliomas are rare tumors causing considerable difficulty in both, diagnosis and treatment. They can be unicentric or multicentric, tend to be locally invasive and, therefore have a high incidence of local recurrence. Histological and immunological phenotype of silent extra-adrenal retroperitoneal paragangliomas is no significant difference with functional ones. In this article, we reviewed the experience in our hospital of this uncommon tumor to highlight the diagnosis and treatment of extra-adrenal paragangliomas.

b) Pathophysiology: Paragangliomas are catecholamine-**secreting** tumors arising from the chromaffin cells of the sympathetic ganglia and are known as extra-adrenal pheochromocytomas. These tumors commonly present with episodic hypertension, tachycardia, headache, and diaphoresis, and can be either benign or malignant.

c) Clinical Perspective: Usually asymptomatic. Rarely can present with chest tightness due to mass effect. More rarely may cause breathlessness or dysphagia due to compression of trachea or esophagus. But in this case patient present with complain of left flank pain.

d) Imaging Perspective: on USG, lobulated solid cystic lesion with evidence of internal vascularity within noted adjacent to inferior pole of left kidney.

On CECT abdomen, Lobulated soft tissue density lesion with subtle calcification with central (cystic component noted in left lumber region at L3-L4 vertebral level, extending from inferior pole of left kidney up to aortic bifurcation which show heterogenous postcontrast enhancement with feeding arterial channels.

Overall histological and immunohistochemistry findings suggestive of extra adrenal Paraganglioma.

e) Treatment options: Surgery remains the mainstay of treatment of extra-adrenal PGLs of retroperitoneum. However, it must be born in mind that the "silent" extra-adrenal PGLs of retroperitoneum is silent ones relatively in the handling of these tumors. Carrying out of physical maneuvers on the tumor and the employment of drugs that free catecholamines can induce hypertensive crises. Once the diagnosis and drug preparation are accomplished, attempt should be made to perform a complete surgical resection. Resection is often challenging as these highly vascular tumors are located near multiple vital blood vessels.

f) Final Diagnosis: extra adrenal paraganglioma

g) Differential diagnosis:
- Schwannoma
- Neurofibroma
- Ganglion neuroma

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