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Original Research Paper

Radiodiagnosis

RETROPERITONEAL PARAGANGLIOMA : A RARE CASE REPORT

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ABSTRACT Extra-adrenal paraganglioma of the retroperitoneum is a very rare neoplasm arising from cells of the primitive neural crest cells. We report a case of an 14-years-old boy, who has been referred to our Radiodiagnosis department, Maharani Laxmi Bai Medical college,Jhansi who presented with acute abdominal pain for 2 days with unexplained hypertension. The CECT Abdomen suggested it to be a retroperitoneal paraganglioma.

KEYWORDS : Paraganglioma, Retroperitoneal, CECT.

INTRODUCTION

Paragangliomas are very rare tumors of sympathetic and parasympathetic paragangliacells. The paragangliomas commonly occur at the carotid body, jugular foramen,middle ear, Aortopulmonary region, Posterior mediastinum, Organ of zuckerkandl in bilateral para-aortic regions of Retroperitoneum more at the origin of inferior mesenteric artery in decreasing order of incidence. Interestingly, the paraganglioma of adrenal medulla is called as phaeochromocytoma. Retroperitoneal paragangliomas are either functional or non functional. Only handful of cases of retroperitoneal paragangliomas are reported in literature so far. We present a very rare case of Retro peritonealparaganglioma.

CASE REPORT:

14 years old male patient was referred from the surgery dept for complaints of acute abdominal pain (Rt. Flank pain) past 2 days.

BP was 140/100 at the time of presentation.

CECT abdomen was done for furthur evaluation.

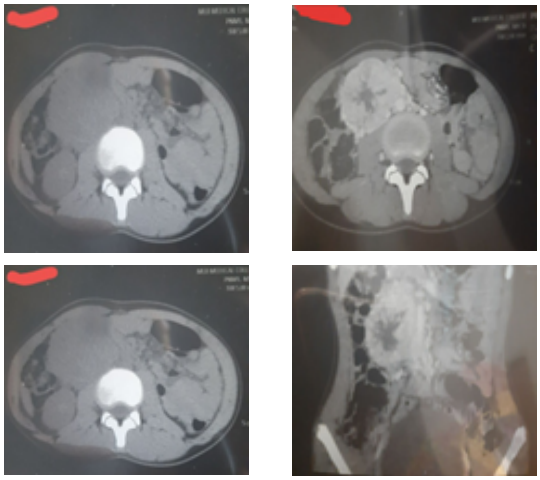

Figure 1

On NCCT, A well definedheterogenous solid mass lesion is noted in the right paravertebral region of size approx. 4x5x6.5 cms and that appears to cause anterior displacement of bowel loops...Suggesting retroperitoneal origin.The lesion is iso to hypodense to adjacent muscle. There is internal hypodense areas suggesting necrosis. No evidence of haemorhhage or calcific foci. IVC appears collapsed. On contrast administration, The arterial phase shows avid enhancement of the mass lesion with internal areas of necrosis....likely a retroperitoneal mass lesion(Neurogenic)

The most likely diagnosis Neurogenic---Paraganglioma (Hyperattenuating/ No calcifications/ Bleeds more commonly)

Differentials Include:

a. Other neurogenic tumors like schwannoma/Neurofibroma (hypoattenuating/ Calcific++)

b. Leiomyosarcoma

Table 1 :

Features	Neurogenictumor	Leiomyosarcoma
Age	Youngagegroup	Oldagegroup
Nature	Benign/Malig	Malignant
Prognosis	Betterprognosis	Poorprognosis
Nature	Solidmass	Solidmass
Degeneration	Cysticdegeneration +++	+
Calcification	Calcification+++	No
Origin	Fromneuralorigin	Fromsmooth muscle,blood vessels
Size	Small	Large
Location	Extravascularalways	ExtraVascular/Intra Vascular
Necrosis	Necrosis+/-	Necrosiscommon
Internalbleed	Haemorrhagewithfluid fluidlevels	Nobleed

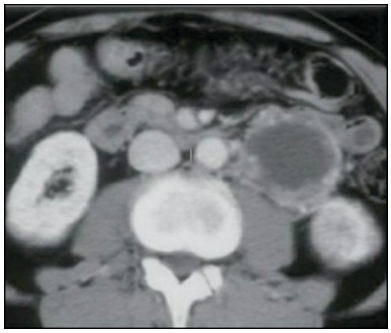

Figure 2: Paraganglioma

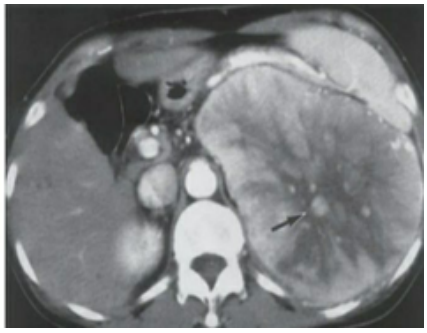

Figure 3:Leiomyosarcoma

DISCUSSION

Retroperitoneal Paraganglioma:

- Associated with NF1,MEN,VHL
- 40 years,Malignant degeneration in younger patients. M=F
- Paraganglia cells-symp-catecholamines.parasymp.
- Retroperitoneum-Organ of Zuckerkandl-Neural crest cells
- MC along Aorta from Inferior Mesenteric Artery origin to bifurcation
- Well defined Vascular solid mass with Intense enhancement,
- bleeds with fluid fluid level
- Malignant feature-internal necrosis
- Punctate calcification+/-
- No light bulb in retroperitoneum
- MIBG-Functional
- Only criteria to tell malignant degeneration- by metastatic disease

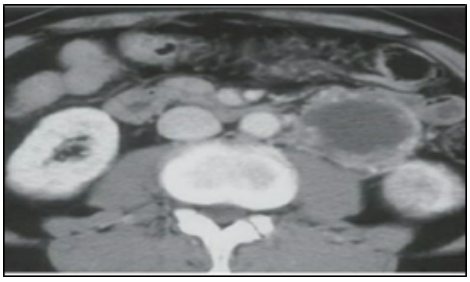

Figure 4: Paraganglioma

Imaging Differentials: Other Neurogenic tumors-Schwannoma, Neurofibroma,Ganglioneuroma and Nerve sheath tumors

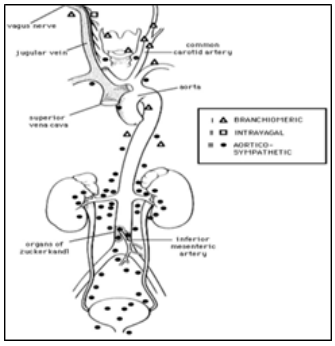

Figure 5

Features	Schwannoma	Neurofibroma
Sex	Females	Males
Number	Single	Multiple
Internalfeature	Calcification++	--
Shape	Round	Dumbell(ifNFinv)
Capsule	Capsulated	Noncapsulated
Cysticchanges	Cysticdegen common-heterogenous	Nocysticdegen-homo enh-Hypoattenuating sincelipidrich
Shape	Roundshaped/dumb belltumor	Dumbellshaped tumorwithwideningof neuralforamina, causingposterior vertebralbody scalloping
Ass.with	NF2,Nomalignant degen	NF1-Malignant degencommon

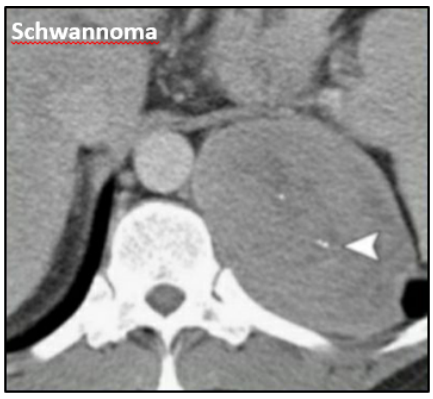

Figure 6

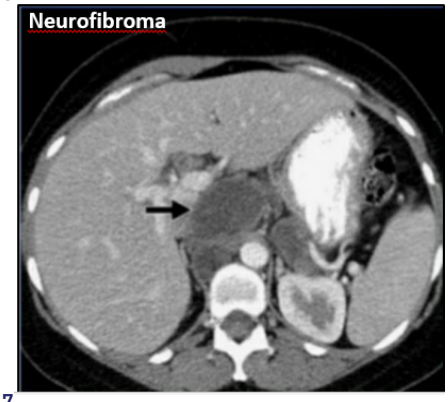

Figure 7

Features	Ganglioneuroma	NST (NerveSheath Tumor)
Attenuation	Large Hypoattenuating	Small, Hypoattenuating
Age	Children	Older
Calcification	MORECALCIFIC FOCI-DISCRETE PUNCTATE Dd-Neuroblastoma-Coarseamorphous calcification	Lesscalcific

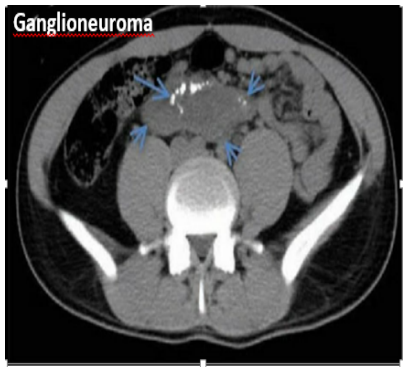

Figure8

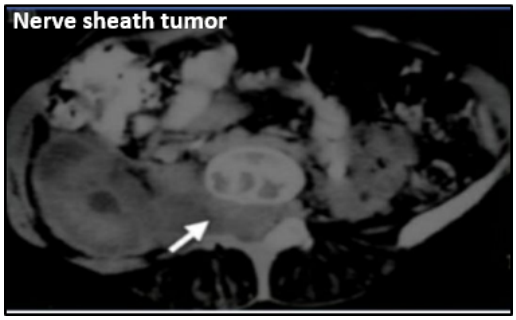

Figure 9

CONCLUSIONS

Paragangliomas are rare tumors with a limited number of cases reported. The localization in the retroperitoneal region is uncommon and is a challenge for surgical resection. Imaging evaluation with CECT plays a very crucial role in the diagnosis of the Retroperitoneal Paragangliomas. Complete surgical resection remains the only curative treatment. Lifetime follow-up with CT imaging is necessary to detect recurrences.

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