Retroperitoneal mass: a case of an extra-adrenal paraganglioma

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Section: Abdominal imaging
Area of Interest: Abdomen
Procedure: Biopsy
Procedure: Diagnostic procedure
Imaging Technique: CT
Special Focus: Cysts Case Type: Clinical Cases
Authors: O'Connor S, Hanson J
Patient: 82 years, female

Clinical History:

An 82-year-old female patient presented with symptomatic congestive cardiac failure. She was found to be anaemic which was investigated. Subsequent imaging showed an incidental retro-peritoneal cystic mass.

Imaging Findings:

Imaging showed an incidental left retro-peritoneal cystic focus. This was separate to the left adrenal. There was layering of debris within the lesion, which measured 7 x 5 cm.

Histology revealed an extra adrenal paraganglioma.

Discussion:

Paraganglioma is a term used to describe any tumour of neural crest – derived endocrine cells or organ [1]. They can arise in any location in the body, though 85%-90% occur in the adrenal medulla and the World Health Organisation classification has reserved the term pheochromocytoma for such tumours. [1, 2] Extra-adrenal paragangliomas account for remaining 15% and tend to occur below the diaphragm, with the majority appearing in the superior para-aortic region between the diaphragm and the lower renal poles. Other sites include the organ of Zuckerkandl, thoracic para-spinal region, bladder and head and neck [2].

They are both catecholamine-producing tumours but whereas pheochromocytomas are known to produce both adrenaline and noradrenaline, extra-adrenal paragangliomas produce noradrenaline and rarely dopamine [3]. Extra-adrenal paragangliomas also carry a higher rate of malignancy then their intra-adrenal counterparts (~20% Vs ~5% for pheochromocytoma [1]) however, figures vary and can range as high as 40% depending on the study [2].

Complete surgical excision is the treatment of choice with lifelong follow up particularly important for extra-adrenal tumours as they are more likely to recur and to metastasise.

Optimal imaging modality for this is difficult to determine due to the rarity of the tumour leading to small study sizes. Both CT and MRI have their advantages and disadvantages, but a review by Sahdev et al. would suggest that perhaps MRI may be more beneficial as it provides greatest variability and can provide additional information when compared to CT for surgical planning [4].

Furthermore, to date there are no morphologic criteria by which benign and malignant tumours can be distinguished making the choice of imaging modality very much dependent on the interpreter experience [5].

Differential Diagnosis List: Histology revealed an extra adrenal paraganglioma, Exophytic pseudocyst, Duplicaion
Final Diagnosis: Histology revealed an extra adrenal paraganglioma

References:


**Description:** Axial CT image showing a left retroperitoneal cystic mass, white arrow. The left renal vein is displaced posteriorly by the cystic lesion. **Origin:** Dr. JM Hanson
Figure 2

Description: Coronal reconstruction. The black arrow is highlighting the tail of the pancreas and the left adrenal gland distant from the epicenter of the retroperitoneal cystic mass. Origin: JM Hanson
Description: Coronal reconstruction, showing the retroperitoneal mass lesion close to the left adrenal gland (white arrow) and pancreas (black arrow). Note, the epicenter is distant to both. Origin: Dr J Hanson
Description: Sagittal reconstruction, showing a fluid level within the retroperitoneal cystic mass, presumed haemorrhage (arrowheads). Note the left kidney (black arrow) and pancreas are separate. 
Origin: Dr. J Hanson
Figure 4

Description: Axial CT image acquired post biopsy, hence the prone position. Note the air-fluid level (white arrow) Origin: Dr J Hanson