Case 14635

Eurorad

Abdominal paraganglioma

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Section: Uroradiology & genital male imaging
Area of Interest: Oncology Abdomen
Procedure: Education
Imaging Technique: Ultrasound
Imaging Technique: Ultrasound-Power Doppler
Imaging Technique: CT
Special Focus: Pathology Neoplasia Case Type: Clinical Cases
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Patient: 32 years, female

Clinical History:

A 32-year-old woman presented to the emergency department complaining of weight loss and the recent appearance of a pulsatile tumefaction at the level of the umbilicus. There were no other relevant clinical aspects.
Imaging Findings:

Ultrasound showed a round, homogeneous and hypoechoic mass, located in the mesogastrium, medially to the inferior vena cava, with sharp borders, demonstrating vascularization at colour Doppler.
On CT the mass appeared to arise from the retroperitoneum, on a paravertebral location, laterally to the abdominal aorta and slightly superiorly to its bifurcation. The mass had round borders and avid and homogeneous enhancement after intravenous contrast administration. There was some compression of the inferior vena cava.
Discussion:

Paraganglioma is a rare neuroendocrine tumour arising from paraganglionic chromaffin cells scattered throughout the body. They can arise from the adrenal medulla, where they are called pheochromocytoma, or from practically every healthy paraganglia: around the carotid body, jugular foramen, middle ear, aorticopulmonary region, posterior mediastinum, and abdominal paraaortic region [1].
Although retroperitoneal paragangliomas can occur anywhere in the paraaortic region, these tumours are frequently found near the origin of the inferior mesenteric artery, where the organs of Zuckerkandl are located. They affect mainly adults in the fourth or fifth decade of life, without sex predilection. Even though most paragangliomas are solitary and arise sporadically, they can be multicentric, and familial occurrence is well recognized [1, 2].
These tumours are functional in more than half of cases, and patients commonly present with symptoms related to excess of secretion of catecholamine, such as palpitations, headache, sweating, and hypertension. For those patients with non-functioning extra-adrenal paragangliomas, the classic clinical manifestation is an insidiously enlarging palpable mass or pain related to the local growth of a mass [2]. In addition, approximately 10% of paragangliomas are clinically silent and detected incidentally at imaging study [3].

On CT they are extremely vascular, usually showing homogeneous enhancement after contrast administration. However they can be large, lobulated and heterogeneous masses due to presence of necrosis, blood and calcification [3, 4]. Smaller tumours are more likely to be homogeneous and sharply marginated compared with the larger ones [3].

MRI demonstrates a hypo- or isointense tumour on T1WI which is markedly hyperintense on T2WI and with a

variable, although often intense, contrast enhancement on T1FS after gadolinium [4]. Internal haemorrhage and cystic components may contribute to a heterogeneous MRI appearance of these tumours [5].
Ideally, the biochemical diagnosis is made before imaging is performed. If there is catecholamine hypersecretion, functional imaging with I123 metaiodobenzylguanidine (MIBG), a catecholamine analogue, can be useful to confirm the diagnosis, identify sites of disease, and evaluate metastases [4].
Paragangliomas are mostly benign with good prognosis, but can be locally invasive and metastasize. Dissemination occurs both lymphatically and haematogenously, with the most common sites of metastasis being the regional lymph nodes, bone, liver, and lung [4].
The possibility for malignant transformation of paragangliomas makes surgical excision the treatment of choice. Radiation therapy has been advocated for patients who are not eligible for surgery or for unresectable tumours. Therapy with radionucleotides may be used for tumours exhibiting uptake on diagnostic scan [4].
Differential Diagnosis List: Retroperitoneal paraganglioma, Schwannoma, Retroperitoneal desmoid tumour

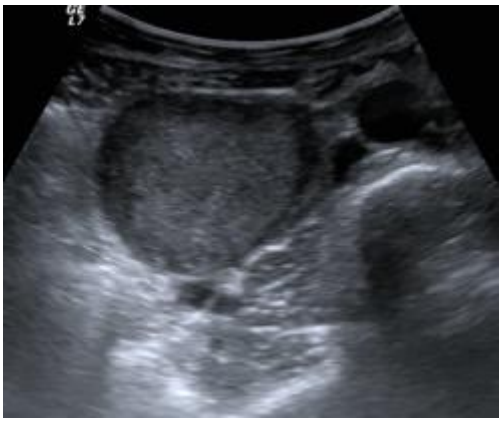
Final Diagnosis: Retroperitoneal paraganglioma

References:

Rha SE1, Byun JY, Jung SE, Chun HJ, Lee HG, Lee JM. (2003) Neurogenic Tumors in the Abdomen: Tumor Types and Imaging Characteristics. Radiographics 23(1):29-43 (PMID: 12533638)
Nishino M et al (2003) Primary Retroperitoneal Neoplasms: CT and MR Imaging Findings with Anatomic and Pathologic Diagnostic Clues. Radiographics 23(1):45-57 (PMID: 12533639)
Lee KY et al (2006) Extraadrenal Paragangliomas of the Body: Imaging Features. American Journal of Radiology 187(2):492-504 (PMID: 16861555)
Brennan C et al (2014) Solid malignant retroperitoneal masses—a pictorial review. Insights Imaging 5(1):53-65 (PMID: 24293303)
Rajia P et al (2011) Imaging of Uncommon Retroperitoneal Masses. Radiographics 31(4):949-76 (PMID:21768233)

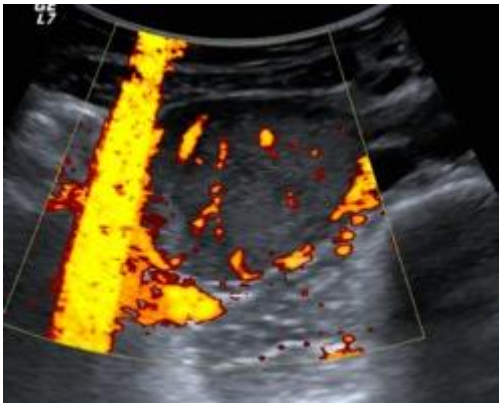
Figure 1

a



Description: Ultrasound shows a round, homogeneous and hypoechoic mass located in the mesograstrium. **Origin:** Department of Radiology, Braga Hospital

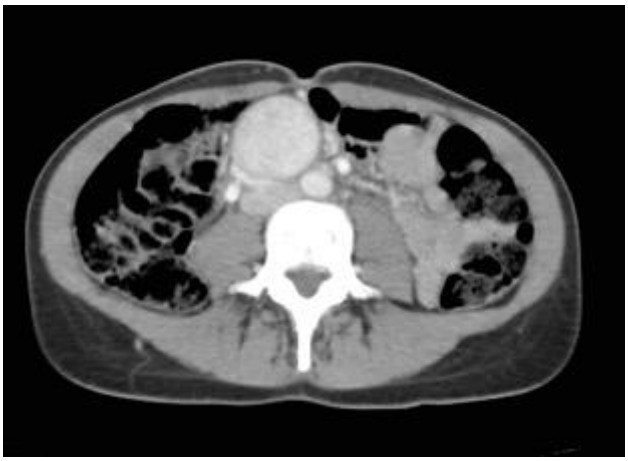
b



Description: On power Doppler, the mass demonstrates clear internal vascularization. **Origin:** Radiology Department Braga Hospital

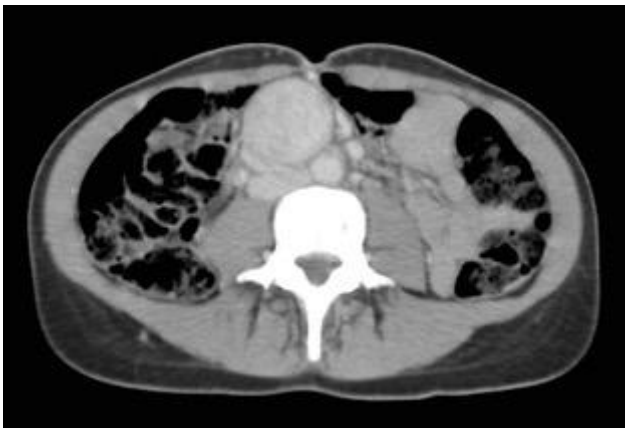
Figure 2

a



Description: CT demonstrates a retroperitoneal mass, located between the aorta and vena cava, with marked enhancement on the late arterial phase. **Origin:** Braga Hospital Department of Radiology

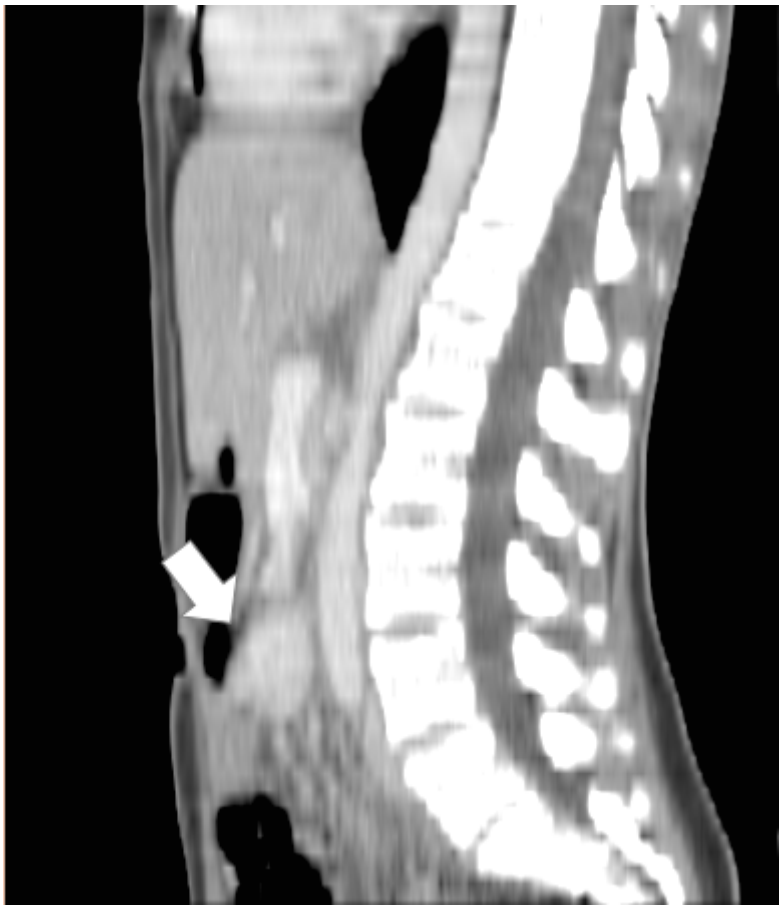
b



Description: On the portal phase the enhancement persists. It is possible to see that the mass doesn't invade any of the adjacent structures, only causes a little bulging of the inferior vena cava. **Origin:** Braga Hospital Radiology Department

Figure 3

a



Description: Sagittal CT confirming the retroperitoneal location of the mass, slighly superiorly to the level of the aortic bifurcation. **Origin:** Braga Hospital Radiology Department