

CLINICAL CASE – TEST YOURSELF

Abdominal Imaging

Incidental pancreatic mass in a female with newly-diagnosed breast cancer

Ekaterini Manavi, Athanasia Mimila, Ekaterini Xinou

Department of Radiology, Theagenio Cancer Hospital, Thessaloniki, Greece

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PART A

A 67-year-old female underwent left lumpectomy for an invasive mucinous breast carcinoma. She had an unremarkable medical history and physical examination revealed only the known palpable mass in the left breast. Laboratory findings were all within normal limits.

Chest and abdomen computed tomography (CT) scan was performed for staging purposes (**Fig. 1**). Due to abnormal findings in the pancreatic region, the patient underwent a multiphasic pancreatic CT (**Figs. 2, 3**) and magnetic resonance imaging (MRI) (**Figs. 4, 5**).

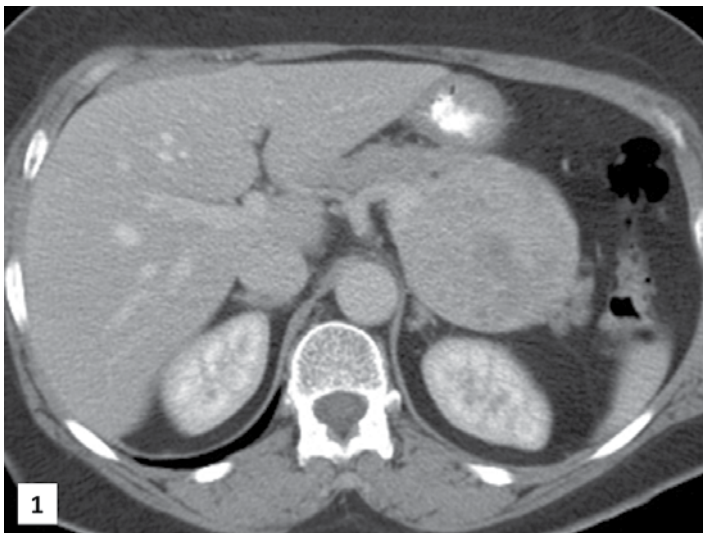


Fig. 1. Axial contrast-enhanced CT image.



CORRESPONDING
AUTHOR,
GUARANTOR

Ekaterini Xinou
Department of Radiology, Theagenio Cancer Hospital, 2 Al. Simeonidi Str, 54007
Thessaloniki, Greece, Email: katxinou@otenet.gr



Fig. 2. Axial contrast-enhanced CT image during the late arterial phase.



Fig. 3. Coronal contrast-enhanced CT image reconstruction during the portal venous phase.

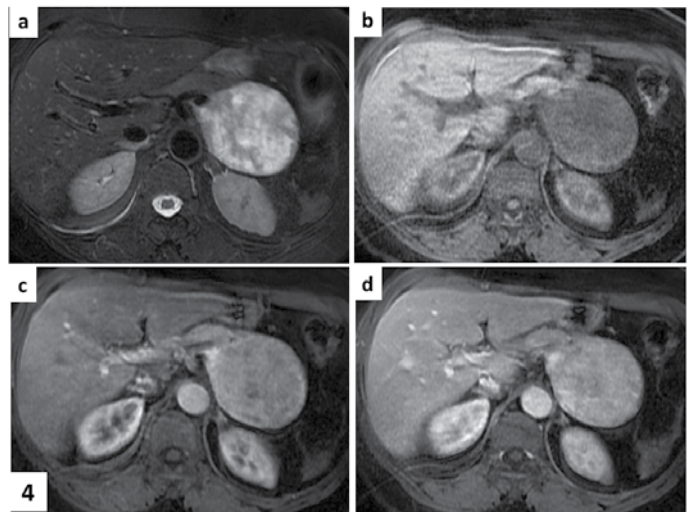


Fig. 4. Dynamic MR. **a.** Axial fat suppressed T2w MR image. **b.** Axial T1w MR image without IV contrast. **c, d.** Axial T1w images with IV contrast (**c:** late arterial phase, **d:** portal phase).

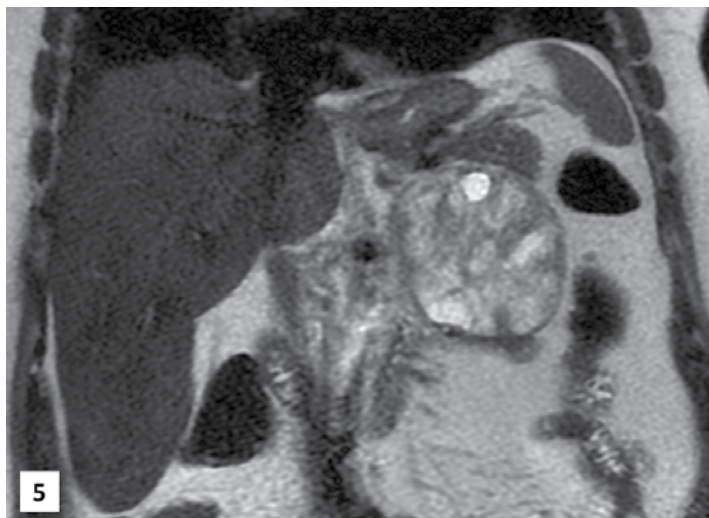


Fig. 5. Coronal T2w MR image.

PART B

Diagnosis: Retroperitoneal paraganglioma

Paragangliomas are rare neuroendocrine tumours arising from the paraganglia, which refer to chromaffin cells of the sympathetic paravertebral ganglia of the thorax, abdomen and pelvis, or of the parasympathetic ganglia located along the glossopharyngeal and vagal nerves in the neck and base of the skull [1, 2]. The adrenal medulla hosts the largest proportion of paraganglia. Paragangliomas located in the adrenal medulla are known as pheochromocytomas and account for 90% of the tumours, whereas when they are located anywhere else they are classified as extra-adrenal paragangliomas. Approximately 70% of sympathetic paragangliomas are intraabdominal, usually in the retroperitoneum, and the remaining 30% are located in the chest [3, 4].

The incidence of extra-adrenal paragangliomas is only 2-8 per million [5]. They occur either sporadically or as part of hereditary syndromes, such as MEN2, von Hippel Lindau, hereditary PGL/pheochromocytoma syndromes and Carney syndrome (extra-adrenal paraganglioma, GIST, pulmonary chondroma) [1, 5]. Extra-adrenal para-

gangliomas are more aggressive than their adrenal counterparts, with metastases occurring in 20-70% of cases, compared to only 2-10% of adrenal pheochromocytomas [4, 5]. Dissemination is lymphatic or haematogenous with the most common site of metastasis being the regional lymph nodes, bone, liver and lung. These tumours are usually locally invasive with a high incidence of local recurrence [2].

Paragangliomas can be divided based on their ability to secrete hormones, mainly catecholamines, into functioning and non-functioning. Functioning paragangliomas secrete norepinephrine and normetanephrine and account for 30-60% of tumours [3]. Catecholamine production may lead in clinical manifestations including palpitation, headache and hypertension [1, 6]. On the contrary, non-functioning paragangliomas tend to be asymptomatic and can be found incidentally, like in our case, or as masses compressing the surrounding organs and causing abdominal pain [6]. Owing to the loose connective tissue of the retroperitoneum, tumours can be very large at presentation [7].

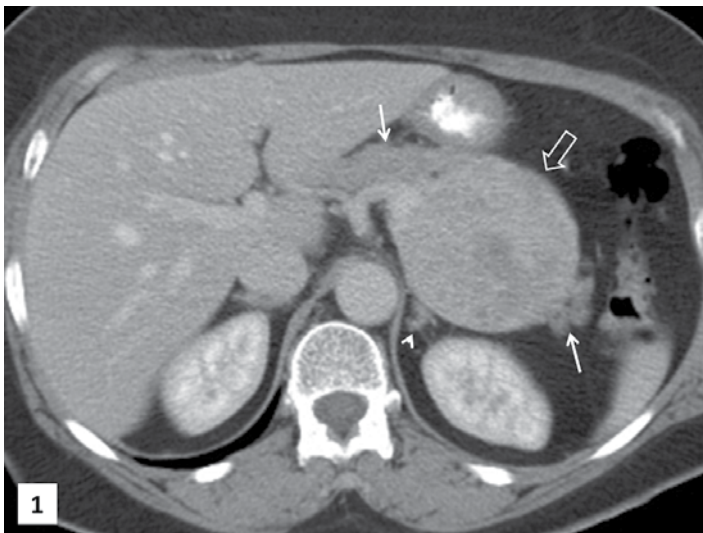


Fig. 1. Axial image of the initial contrast-enhanced CT scan showing a large enhancing tumour (open arrow) in the pancreatic tail with clear fat plane with the left adrenal gland (arrowhead) and the pancreas (arrows).



KEY WORDS

retroperitoneal mass; paraganglioma; CT/diagnosis; MR imaging/diagnosis



Fig. 2: Axial image of the subsequently performed multiphase pancreatic CT scan showing the intense inhomogeneous enhancement of the tumour (open arrow) during the late arterial phase and the areas of necrosis; pancreas (arrows), left adrenal gland (arrowhead).



Fig. 3. Coronal reconstruction of the same multiphase pancreatic CT during the portal venous phase showing the clear borders and the extrapancreatic location of the tumour (open arrow), which dislocates cranially the pancreatic tail (arrows). Note the more pronounced enhancement of the tumoural capsule (open arrow) and the absence of pancreatic duct dilatation (arrows); splenic vein (arrowhead).

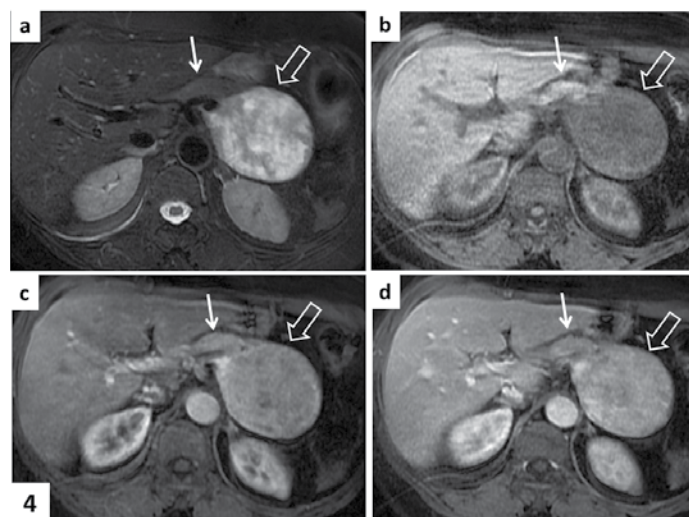


Fig. 4. Dynamic MR. **a.** Axial fat suppressed T2w MR image showing the inhomogeneous, high signal intensity of the tumour (open arrow); pancreas (arrow). **b-d.** Axial T1w images before (**b**) and after IV contrast (**c, d**) showing the intense inhomogeneous enhancement of the tumour during the late arterial phase (**c**), which persists in the portal phase (**d**); tumour (open arrow); pancreas (arrow).

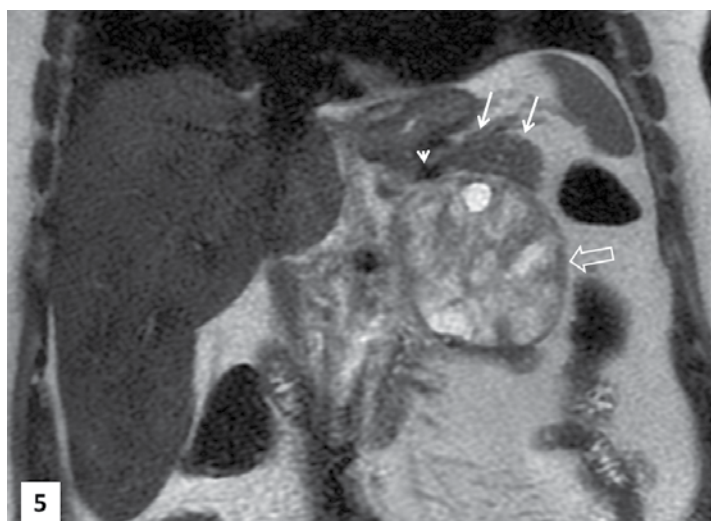


Fig. 5. Coronal T2w MR image showing the extrapancreatic location of the well-circumscribed tumour (open arrow), which dislocates cranially the pancreatic tail (arrows). Note the absence of pancreatic duct dilatation (arrows); splenic vein (arrowhead).

If a retroperitoneal mass is disclosed on imaging and suspicion of a neuroendocrine tumour is raised, the diagnosis of a paraganglioma is usually established with the presence of elevated levels of vanillylmandelic acid and metanephrine in 24-hour urine collections [4]. The above mentioned catecholamine metabolites are increased in over 90% of paraganglioma cases [6].

Retroperitoneal paragangliomas are classically associated with the organ of Zuckerkandl, the chromaffin paraortic tissue between the origin of the inferior mesenteric artery and the aortic bifurcation [8]. Paragangliomas arising from the pancreas are extremely rare and it has not been clarified whether they represent an extension of a retroperitoneal tumour or they derive from ectopic paraganglia located in the pancreas. Of the 22 cases reported in the literature, the vast majority concerned the pancreatic head [2].

On unenhanced CT, paragangliomas have typically density greater than 10 Hounsfield units (HU) [4]. On contrast-enhanced CT, they show avid enhancement due to a rich capillary network and delayed wash-out. Internal haemorrhage, necrosis and calcifications are usually seen within the tumours. CT exceeds MRI in spatial resolution and has a sensitivity of around 90% for identifying extra-adrenal paragangliomas [1, 7]. MRI allows further evaluation of the lesions, due to superior tissue characterisation. Tumours are generally characterised by low T1 and bright T2 signal intensities [5]. Nevertheless, T2-weighted images demonstrate the characteristic uniform high-signal intensity ("light bulb") only in about 80% of cases, because the presence of haemorrhage may reduce signal intensity [7, 8]. Haemorrhagic areas can be bright on T1-weighted images. Fluid-fluid levels can also be seen in some cases [7]. MRI is more sensitive than CT in detecting extra-adrenal tumours [3]. Scintigraphy with 123-I labelled MIBG offers superior specificity than CT and MRI (90-100%) in localising extra-adrenal disease and metastases [5, 9].

On histopathology, paragangliomas are diagnosed by their highly vascular appearance, with chief cells and sustentacular cells arranged in clusters called "Zellballen" [8, 9]. This histopathologic pattern was also revealed in our case with presence of necrotic areas, correlating with the radiological diagnosis of an inhomogeneously hyperenhancing neuroendocrine tumour. Chief cells were positive

for neuroendocrine markers (synaptophysin, NSE, chromogranin), and sustentacular cells were positive for S-100 protein.

Surgical resection remains the mainstay of treatment of retroperitoneal extra-adrenal paragangliomas [5, 9]. Total excision is often difficult, as these tumours are frequently located adjacent to vital blood vessels. If a mass is unresectable, radiotherapy, embolism or chemotherapy may be indicated to reduce the size [9].

In our patient the incidentally depicted hyperenhancing mass was initially thought to be intrapancreatic, with the most probable diagnosis being neuroendocrine tumour, since solitary pancreatic metastasis from breast cancer is an extremely rare entity (Fig. 1) [10]. No dilatation of the pancreatic duct or encasement of the vasculature by the mass was depicted, and there was no evidence of metastatic disease. Multiphasic CT with pancreatic protocol and dynamic abdominal MRI, which were performed subsequently, revealed in the coronal reconstructions that the tumour was not located in the pancreatic tail, but it was displacing it cranially and ventrally (Figs. 3, 5). This finding changed the differential diagnosis which now included hyperenhancing retroperitoneal tumours such as metastatic lymphadenopathy, sarcomas and paragangliomas.

Tumour location near the aorta, high signal intensity in T2-weighted images and tumoural necrotic changes were the key imaging findings suggesting the correct diagnosis of extra-adrenal paraganglioma (Figs. 2-5). Subsequent CT-guided biopsy confirmed the diagnosis and surgery was then decided. Intraoperatively, the tumour was well-encapsulated and was easily excised without needing to perform distal pancreatectomy.

In conclusion, in cases of large tumours in the pancreatic region where true location is difficult to ascertain, coronal and sagittal reconstructions can be helpful in distinguishing intrapancreatic from extrapancreatic tumours and should always be scrutinised. When faced with an extra-adrenal retroperitoneal hyperenhancing tumour with high T2 signal intensity and necrotic areas in close vicinity with the aorta, radiologists should include the paraganglioma in the differential diagnosis. **R**

Conflict of interest

The authors declared no conflicts of interest.

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