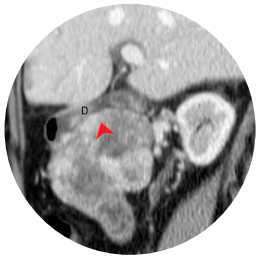

Pancreatic paraganglioma a differential diagnosis of pancreatic mass. Case report

Paraganglioma pancreático, un diagnóstico diferencial de masa pancreática. Presentación de caso

Manuela Laguna Kirof[1]
Víctor Nebil Larrañaga[2]
María De Vedia[2]
Nicolás Roccatagliata[3]
Javier Vallejos[2]

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Summary

Paraganglioma is a rare neoplasm that arises from the chromaffin cells of the autonomic nervous system. Paragangliomas that originate in the pancreas are even rarer with a very low rate of malignancy. Given the small number of cases documented so far, few radiologic characteristics are known about them and they are usually incidental findings in routine imaging studies. The vast majority of pancreatic paragangliomas do not secrete catecholamines and are called "non-functioning" and can cause symptoms such as abdominal pain or be asymptomatic. There are only two cases of catecholamine-secreting or "functioning" paragangliomas documented to date, presenting with headache, hypertension, and palpitations. Pancreatic paragangliomas reported so far have occurred in multiple locations in the pancreas, including the head, body, and tail. The risk of malignant transformation of paragangliomas makes surgical resection the treatment of choice, and aggressive surgery and close postoperative follow-up are mandatory to achieve disease-free survival. In this study we present the case of an 83-year-old patient with a diagnosis of non-functioning paraganglioma confirmed by pathological anatomy, located in the cephalic portion of the pancreas. We also review relevant literature.

Resumen

El paraganglioma es una neoplasia poco frecuente que surge de las células cromafines del sistema nervioso autónomo. Su localización en el páncreas es aún más rara, con una muy baja tasa de malignidad. Dada la escasa cantidad de casos documentados hasta el momento, se conocen pocas características radiológicas de los mismos y suelen ser hallazgos incidentales en estudios de rutina. La gran mayoría de los paragangliomas pancreáticos no secretan catecolaminas y son denominados "no funcionantes", aunque pueden provocar síntomas como dolor abdominal o ser asintomáticos. Hasta donde conocemos, hay solo dos casos de paragangliomas secretores de catecolaminas o "funcionantes" documentado, con cefalea, hipertensión arterial y palpitaciones como forma de presentación. Los paragangliomas pancreáticos descritos hasta el momento se presentaron en diferentes localizaciones del páncreas, incluidos el cuerpo, la cabeza y la cola. El riesgo de transformación maligna de los mismos hace que la resección quirúrgica sea el tratamiento de elección. La cirugía agresiva y el seguimiento posoperatorio estrecho son obligatorios para lograr una supervivencia libre de enfermedad. En este trabajo se presenta el caso de una paciente de 83 años de edad con diagnóstico de paraganglioma no funcionante confirmado mediante anatomía patológica, localizado en la porción cefálica del páncreas. También se hizo la revisión de literatura pertinente.

[1]Second year resident physician, Diagnostic Imaging, Centro de Educación Médica e Investigaciones Clínicas (CEMIC). Ciudad Autónoma de Buenos Aires, Argentina.

[2]Specialist in Diagnostic Imaging, Centro de Educación Médica e Investigaciones Clínicas (CEMIC). Ciudad Autónoma de Buenos Aires, Argentina.

[3]Specialist in Diagnostic Imaging, First Year Fellowship, Abominopelvic Imaging,Hospital Británico. Ciudad Autónoma de Buenos Aires, Argentina.

Introduction

Paraganglioma is a rare neoplasm that originates in the neural crest and affects the ganglia of the sympathetic trunk, as well as the celiac, renal, adrenal, aortic and hypogastric pleural ganglia.

The incidence is approximately 1 in every 2,000,000 adults, with a malignancy rate of approximately 10% and a survival rate of 50% at 5 years (1). When they do not metastasize they are considered benign lesions. Only 31 cases of pancreatic paragangliomas have been documented (1) with an average age at presentation of 42 to 85 years and a male to female incidence rate of 1:2.

Computed axial tomography (CT) findings, as described in the literature, show masses of equal or low density with respect to the underlying pancreatic parenchyma, with enhancement in the arterial phase. They may also present cystic changes and areas of necrosis in their interior (2).

The preoperative diagnosis is often challenging, especially in cases of non-functioning tumors, since it is difficult to differentiate it radiologically from other pancreatic neoplasms with arterial enhancement, such

as neuroendocrine tumors or vascular lesions (1). Surgical resection is necessary for the evaluation and histological confirmation of these tumors (3).

Clinical Case

An 83-year-old female patient with a history of cigarette smoking up to 15 years ago and a history of dyslipidemia. She was admitted to the emergency department for presenting tarry stools and presyncopal episode. A CT scan of the abdomen and pelvis was performed, with oral and intravenous contrast medium, showing a solid lesion in the head and uncinate process of the pancreas, with heterogeneous enhancement in arterial phase after administration of intravenous contrast medium (Figure 1). The lesion was in contact with the second and third portion of the duodenum (figure 2), and extended towards the pancreaticoduodenal sulcus (figure 3). Based on these findings, a cephalic duodenopancreatectomy was performed, which showed, according to the histopathological report, a neoplasm formed by tapered cells with irregular nuclei and prominent nucleoli, arranged in nests (Zellballen) surrounded by a myxoid type stroma with congestive vascular structures and areas of recent hemorrhage, compatible with pancreatic paraganglioma.

Discussion

Although the best known location of paragangliomas is the adrenal medulla, where they are called pheochromocytomas, approximately 5% to 10% develop in extra-adrenal sites, 85% of them retroperitoneal and between the origin of the inferior mesenteric artery and the aortic bifurcation (2).

Paragangliomas located within the pancreas are extremely rare, with few cases documented to date (3).

These tumors can be "functioning" or "non-functioning". The former usually manifest themselves by secretion of catecholamines and cause hypertension (persistent or paroxysmal), palpitations and excessive generalized sweating, or they may be asymptomatic. Non-functioning paragangliomas may remain asymptomatic, even when there is a considerable mass, as seen in this case, and can easily mimic other retroperitoneal masses on abdominal imaging.

The ultrasound findings of pancreatic paraganglioma are: an ill-defined, heterogeneous, predominantly hypoechoic mass that may present internal anechoic areas and with abundant vascularization on Doppler scanning.

In CT it usually appears as a solid mass of medium or low density, well delimited, with intense enhancement after the administration of intravenous contrast medium, with avascular areas due to the presence of necrosis or cystic changes (figure 1) (3); calcifications are an infrequent finding.

Magnetic resonance imaging (MRI) is more sensitive than CT to visualize extra-adrenal tumors. The usual appearance is that of an oval or rounded mass, with low signal in T1-weighted sequences and high signal in T2-weighted sequences compared to the adjacent normal pancreatic parenchyma. Its hypervascularization is responsible for the intense enhancement in arterial phase after intravenous administration of contrast medium (4).

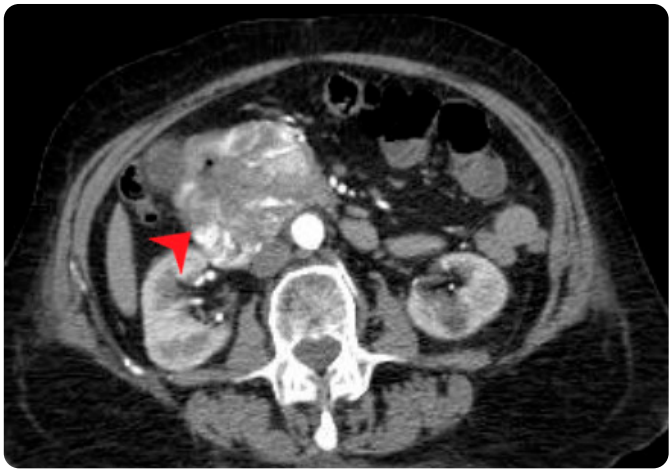

Figure 1. CT with intravenous contrast medium in arterial phase, axial: solid retroperitoneal image, in head and uncinate process of the pancreas, with heterogeneous enhancement.

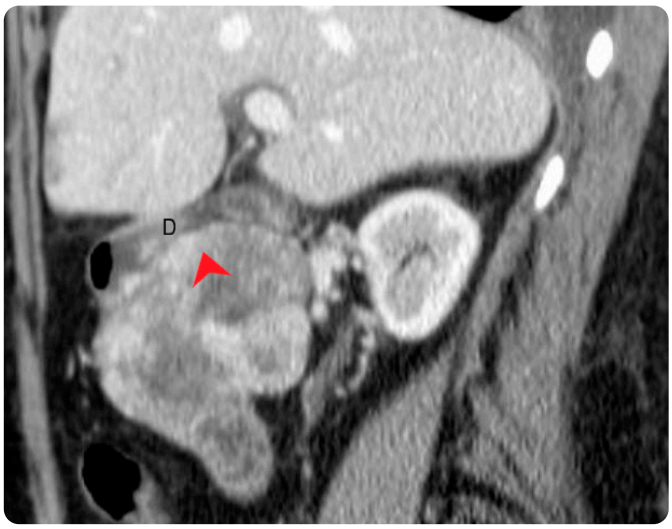

Figure 2. CT with intravenous contrast medium in venous phase, sagittal: solid lesion (red arrow) in intimate contact with the second and third portion of the duodenum, without evidence of infiltration. D: duodenum.

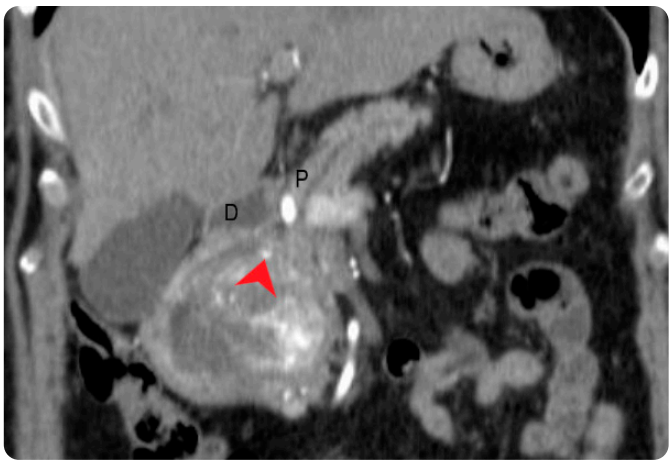

Figure 3. Abdominal CT, coronal: heterogeneous solid mass expanding towards the pancreaticoduodenal sulcus (red arrow). P: pancreas. D: duodenum.

Since the tumors are non-functioning and the intrapancreatic location is variable, the preoperative diagnosis of paraganglioma is speculative and the primary pancreatic malignant tumor (adenocarcinoma) is a feasible differential diagnosis. The difference is that the latter usually appear on MRI as poorly defined masses, with low signal in T1 sequences with little enhancement in the pancreatic phase and, sometimes, enhancement in the late phases. In turn, they cause dilatation of the biliary and pancreatic ducts with retroperitoneal involvement (1). It should be noted that dilatation of the bile duct is infrequent in cases of pancreatic paragangliomas, even when they are located in the cephalic portion of the pancreas (3).

These hypervascular tumors are also often misdiagnosed as pancreatic neuroendocrine tumors (pNET) which can also be functioning or non-functioning depending on whether they present clinical and laboratory findings (5). Functioning pNETs are usually small in size, typically enhance intentionally during the arterial phase and are maintained with slight enhancement during the portal and late venous phases. Large non-functioning pNETs may undergo necrosis and degenerative changes and show heterogeneous enhancement after administration of intravenous contrast medium (6). As a consequence, it is difficult to differentiate them radiologically from paragangliomas, so pathologic diagnosis is usually necessary (7).

Other less likely differential diagnoses are: pseudopapillary solid tumor, represented by a large encapsulated solid-cystic mass and vascular lesions such as aneurysms or pseudoaneurysms. However, when lesions are sufficiently large, Doppler ultrasound can differentiate a purely vascular lesion from a paraganglioma or a neuroendocrine tumor (4).

Histopathological findings can confirm the diagnosis of paraganglioma by microscopic evidence of the classic Zellballen pattern formed by well-developed polygonal chromaffin cells surrounded by an intermediate fibrovascular stroma and peripheral sustentacular cells (8).

Because paragangliomas have metastatic potential to the liver, bone and lymph nodes, annual lifelong follow-up is recommended.

Laparoscopic resection for small non-invasive tumors or laparotomy for large tumors is the curative therapy of choice for localized tumors.

Conclusion

In conclusion, despite being an extremely infrequent entity and with such variable biological behavior, paragangliomas should be considered as a differential diagnosis of a hypervascular pancreatic mass, especially in those cases in which enhancement is observed with the administration of contrast medium. Knowing their radiological characteristics will allow us to reach an earlier diagnosis in order to choose an adequate therapeutic approach and improve the survival rate.

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Correspondence

Manuela Laguna Kirof
Galván 4102
Buenos Aires, Argentina
manuelalaguna17@gmail.com

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