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Pancreatic Gangliocytic Paraganglioma – CEUS Appearance

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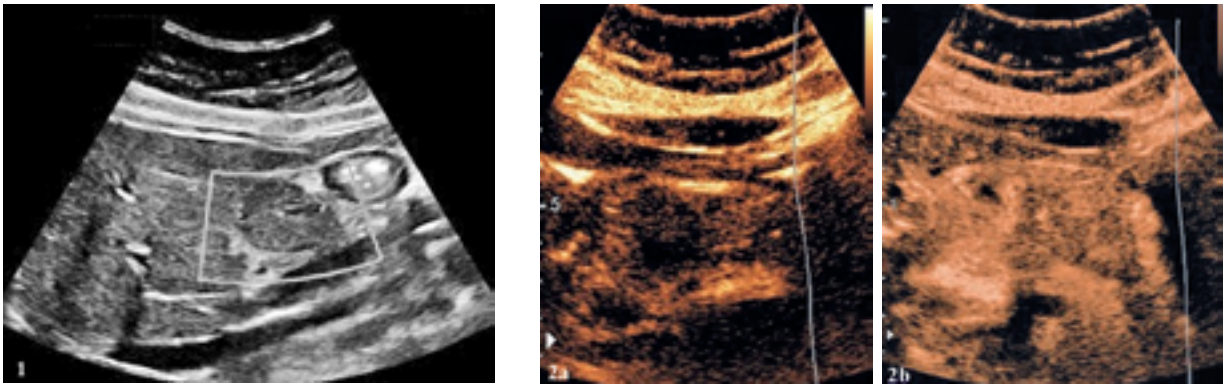

Gangliocytic paraganglioma is a rare neuroendocrine tumour (NET), first described by Dahl et al. in 1957 [1], located in the second portion of the duodenum, in the periampullary region, and containing both paraganglioma and ganglioneuroma structures.

A 53-year-old female patient was admitted for dyspeptic symptoms. Abdominal ultrasound revealed an encapsulated hypoechoic, inhomogeneous tumor in the pancreatic isthmus (Fig 1). The mass did not invade the adjacent structures and did not invade the main biliary or pancreatic ducts. At contrast-enhanced ultrasound (CEUS), this mass intensely captured the contrast agent in the arterial phase, with a few small areas lacking a vascular signal. This feature is highlighted in Fig. 2 showing the pancreatic mass on the basal time (Fig. 2a) with intense arterial enhancement (Fig. 2b). Very early in the arterial phase (second 7), the contrast agent appeared inside this tumor, indicating a rich arterial vascularization. The capture was slightly inconsistent due to some necrotic or cystic areas. All of these elements suggested a NET. The differential diagnosis for hypervascularized pancreatic lesions included hypervascularized metastases (especially of renal cancer), an intra-pancreatic accessory spleen (especially at the tail) or a predominantly serous cystadenoma.

A central pancreatectomy was performed. The histopathological examination revealed a 35/25/25 pancreatic mass, containing three components: epithelial cells, fusiform cells and ganglion cells and established the diagnosis of pancreatic gangliocytic paraganglioma. The mitotic index was 1-2 mitoses/10 HPF without atypical mitosis.

Gangliocytic paraganglioma is one of the four main types of duodenal NETs, along with the gastrinoma, somatostatinoma and enterochromaffin tumour [2]. It is considered a benign tumor, although a malignant potential has been found in 5-7% of the cases [3, 4]. It is well known that the NET tumors are highly vascularized tumors and that CEUS is a valid non-invasive and accurate imaging modality for quantifying tumor vascularity [5]. Gangliocytic paraganglioma has similar CEUS features as other NET tumors and should be considered in the differential diagnosis of hypervascularized lesions with pancreatic location other than the periampullary region, as revealed by our case.

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Conflicts of interest: None to declare.

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