

Conflict of Interests

The authors have no conflict of interests to declare.

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Diagnostic/Therapeutic Management of Pancreatic Neuroendocrine Tumours Associated with MEN 1 Syndrome[☆]



Manejo diagnóstico/terapéutico en los tumores neuroendocrinos pancreáticos asociados a MEN 1

As described by Wermer,¹ multiple endocrine neoplasia type 1 (MEN 1) syndrome is a disorder with a prevalence of 2/100 000 inhabitants. It is characterised by the association of parathyroid adenomas, gastroenterohepatic neuroendocrine tumours and pituitary adenomas,² with a predisposition towards other lesions, such as adrenal and carcinoid tumours, facial angiofibromas, meningiomas, etc.³

Pancreatic neuroendocrine tumours (PNET) represent 3% of the lesions in said location. Most are sporadic but can appear in endocrinopathies such as MEN 1.

The patient is a 55-year-old woman with a genetic diagnosis of MEN 1 after having undergone surgery at the age of 17 due to microprolactinoma and at 26 for primary

hyperparathyroidism related with parathyroid hyperplasia. Follow-up included annual hormone determinations (calcium, PTH, glucose, insulin, glucagon, IGF-1, prolactin and chromogranin A) and cervical/abdominal ultrasound every 3 years, with no abnormal findings for 20 years.

After inconclusive findings on the last ultrasound, magnetic resonance imaging (MRI) was used at the next follow-up, which detected a retroperitoneal mass measuring 16 cm×13 cm×10 cm in contact with the body/tail of the pancreas and left kidney (Fig. 1). With a suspected diagnosis of non-functioning PNET, we decided to operate and performed *en bloc* resection including the tail of the pancreas, spleen, left kidney and descending colon due to infiltration (Fig. 2). The histopathology study reported a

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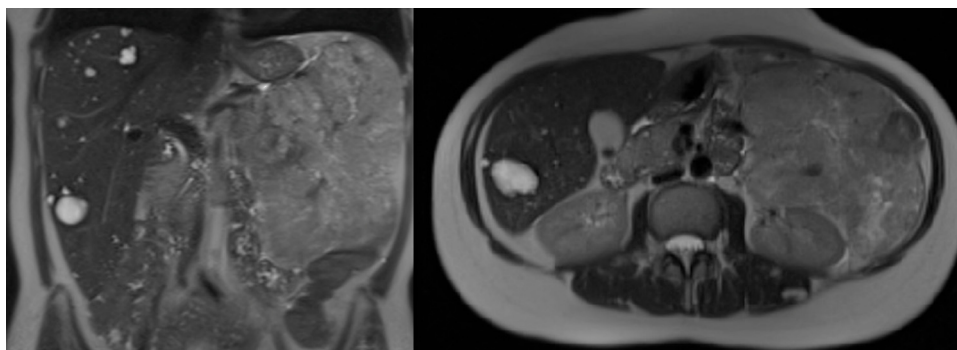


Fig. 1 – Abdominal MRI: retroperitoneal mass in close contact with the body and tail of the pancreas, and left kidney.

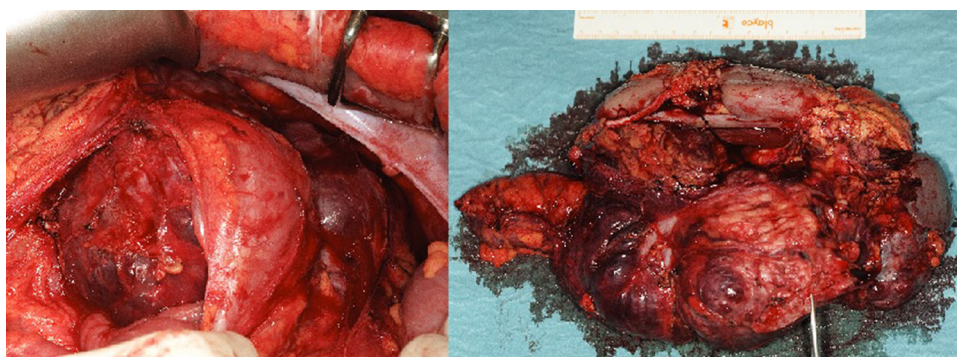


Fig. 2 – Macroscopic images of the procedure; en bloc resection of the tumour.

neuroendocrine tumour (22 cm×16 cm×7 cm) that was moderately differentiated and pancreatic in origin, which surpassed the pancreatic limits (pT3); resected margins were disease free.

Twelve months later, the patient has received no adjuvant treatment and is disease free (octreotide study showed no evidence of lesions).

MEN 1 is an autosomal dominant genetic disorder, although 10% are associated with *de novo* mutations.³ The most frequent genetic alteration is in the MEN 1 gene (chromosome 11), with a penetrance greater than 95% at the age of 40.⁴

Parathyroid adenomas are the most frequent manifestation, with a penetrance of almost 100% at ages 40–50, and typically the earliest, with a presentation some 30 years before sporadic cases.²

Pituitary adenomas present an incidence of 15%–50%, and prolactinomas are the most frequent. They tend to be larger, more aggressive and less responsive to treatment, although the prevalence of carcinoma does not increase.³

Associated PNET usually appear earlier, are multiple and tend to degenerate towards malignancy.⁵

Traditionally, gastrinomas with Zollinger-Ellison syndrome were more frequent (40%).⁶ Advances in imaging studies has resulted in non-functioning tumours being the most frequent in the pancreatic-duodenal region (30%–80%), with a penetrance of 35% at 50 years. More than 70% are not truly non-functioning; instead, they secrete substances like pancreatic polypeptide, enolase or neurotensin,⁷ but entail no clinical repercussions.

As they are not associated with any clinical syndromes and the associated symptoms are vague, diagnosis is made in

advanced stages and at sizes larger than 5 cm in 70% of cases; metastasis is found at the time of diagnosis in 60% of patients.⁷

Indications for surgery are based on tumour growth or size >2 cm; at smaller sizes, no benefits are seen in survival.^{8,9} In the management of lesions measuring 1–2 cm, due to the morbidity and mortality associated with pancreatic surgery and the appearance of new lesions in the remaining pancreas,^{3,6} there is no consensus about the optimal moment for surgery or its extension.

As for the screening of PNET associated with MEN 1, there is no standard protocol, but it should always include an annual biochemical screening (calcium, PTH, prolactin, glucose, insulin and chromogranin A) and variable imaging studies. In some cases, endoscopic ultrasound, thoracoabdominal CT (if there is a history of pancreatic surgery or thymic carcinoid tumour) and somatostatin receptor scintigraphy (if malignant PNET) are recommended every 3 years.¹⁰ Meanwhile, others propose the use of imaging studies every 3–5 years² or annually,³ always depending on the patient's wishes. At our hospital and based on this case, biochemical determinations are currently done annually and abdominal MRI is done every 3 years.

Survival is shorter in this population group compared to a population that is similar in age, sex and other demographic characteristics, with a probability of death of 50% at the age of 50 if not treated. PNET are the main prognostic indicators in MEN 1 and their malignisation is the main cause of death.³ Screening has not been related with increased survival rates.¹⁰

Sizes larger than 3 cm and the presence of metastasis are related with prognosis, and diameters of 1.5–2 cm are related with increased risk of malignisation.^{5,6}

Therefore, our case is relevant due to the presence of a large, non-functioning PNET without metastasis at diagnosis, which resulted in a certain improvement in the prognosis of the patient. We also emphasise the need to standardise the radiological follow-up in these patients.

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Celiac Artery Stenosis due to Median Arcuate Ligament Compression: A Risk Factor in Pancreaticoduodenectomy?^{☆,☆☆}

Estenosis del tronco celíaco por el ligamento arcuato, ¿un riesgo en la duodenopancreatectomía cefálica?

Median arcuate ligament syndrome is caused by compression at the origin of the celiac trunk by the arcuate ligament, a fibrous band that joins the 2 crura of the diaphragm. Although the stenosis is usually asymptomatic, when it interrupts the pancreaticoduodenal arterial arcades (as in pancreaticoduodenectomy), it may lead to visceral ischaemia.

We present the case of a 74-year-old man who was admitted for further study of epigastric pain, general malaise and jaundice. Work-up showed a total Br of 5.50 and GPT of 615. Abdominal

ultrasound reported dilatation of the intrahepatic bile duct and the common bile duct of 15 mm, with no observable cause.

CT demonstrated a mass in the uncinate process of the pancreas, which caused an important dilatation of the bile duct and main pancreatic duct. Results of endoscopic ultrasound-guided FNA were compatible with adenocarcinoma, and it was necessary to insert a plastic stent by ERCP.

Likewise, an important amount of stenosis was observed at the origin of the celiac trunk (Fig. 1) with post-stenotic

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