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FROM ULCER DISEASE TO MEN1: A PANCREATIC NEUROENDOCRINE TUMOR CASE

Author: Zoubida Behourah1
Department of Medical Oncology
Faculty of Medicine, Oran
Oran, Algeria
zbelhamza@yahoo.fr

Co-Authors: Malika Ahed Messaoud1, Amina Bensetti-Houari2, Karima Hamidatou1, Malika Lechar1, Ahlem Megaiz1, Soumeya Ghazli1, Aicha Rabah1, Mohamed Yamouni1, Khadidja Belkharoubi2, Abdelkader Bousahba1

1 Department of Medical Oncology
2 Surgery department

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ABSTRACT

Introduction: Multiple endocrine neoplasia type 1 (MEN1) is a rare autosomal dominant tumor syndrome caused by inactivating mutations in the MEN1 gene. It is characterized by the development of primary hyperparathyroidism (PHPT), pancreatic neuroendocrine tumors (pNETs), and pituitary adenomas. Early diagnosis is essential for implementing appropriate surveillance and management.

Case Presentation: A 34-year-old woman with recurrent duodenal ulcers was found to have multiple pancreatic neuroendocrine tumors, a prolactin-secreting pituitary adenoma, and primary hyperparathyroidism. Imaging and histopathology confirmed a well-differentiated pancreatic neuroendocrine tumor without metastases. She was managed with proton pump inhibitors, somatostatin analogs, dopamine agonist therapy, and surgical evaluation for hyperparathyroidism.

This case highlights the classical MEN1 triad and the importance of comprehensive endocrine and genetic assessment for diagnosis and management.

Introduction:

Multiple endocrine neoplasia type 1 (MEN1) is a hereditary tumor syndrome caused by mutations in the MEN1 tumor suppressor gene located on chromosome 11q13, encoding menin, a nuclear protein that regulates cell proliferation, transcription, and genome stability [1]. MEN1 is inherited in an autosomal dominant pattern with high penetrance; over 90% of mutation carriers develop clinical manifestations by age 50 [1,2].

Clinically, MEN1 is defined by the presence of at least two of the three major endocrine manifestations: primary hyperparathyroidism (PHPT), pancreatic neuroendocrine tumors (pNETs), and pituitary adenomas [1,2]. PHPT is the most frequent and earliest manifestation, often due to multiglandular parathyroid hyperplasia [2,3]. Pancreatic neuroendocrine tumors, especially gastrinomas, are the leading cause of MEN1-related morbidity and mortality [3,4]. Pituitary adenomas, most commonly prolactinomas, tend to present earlier and may behave more aggressively than sporadic tumors [1,2].

Early recognition of MEN1 is essential, as timely diagnosis allows implementation of appropriate surveillance strategies, optimization of therapeutic interventions, and genetic counseling with screening of at-risk family members. This case illustrates the diagnostic value of multimodal imaging in revealing MEN1 and highlights the importance of a systematic endocrine evaluation in young patients presenting with pancreatic neuroendocrine tumors.

Case Presentation: A 34-year-old woman with a long-standing history of recurrent duodenal ulcer disease was referred in February 2022, for further evaluation of persistent epigastric pain despite medical treatment. There was no known family history of endocrine or hereditary disorders.

Initial abdominal ultrasound revealed a 7 mm hypoechoic lesion in the tail of the pancreas, without pancreatic duct dilation. Contrast-enhanced computed tomography demonstrated a hypervascular lesion in the pancreatic tail with intense arterial enhancement and delayed washout, suggestive of a pancreatic neuroendocrine tumor (pNET). Magnetic resonance imaging confirmed a hypervascular lesion 12mm that was hypointense on T1-weighted images, hyperintense on T2-weighted sequences, and demonstrated restricted diffusion on diffusion-weighted imaging (Figure 1). Additionally, imaging revealed right-sided ureteropyelocaliceal dilation due to an obstruction at the level of the proximal ureter.

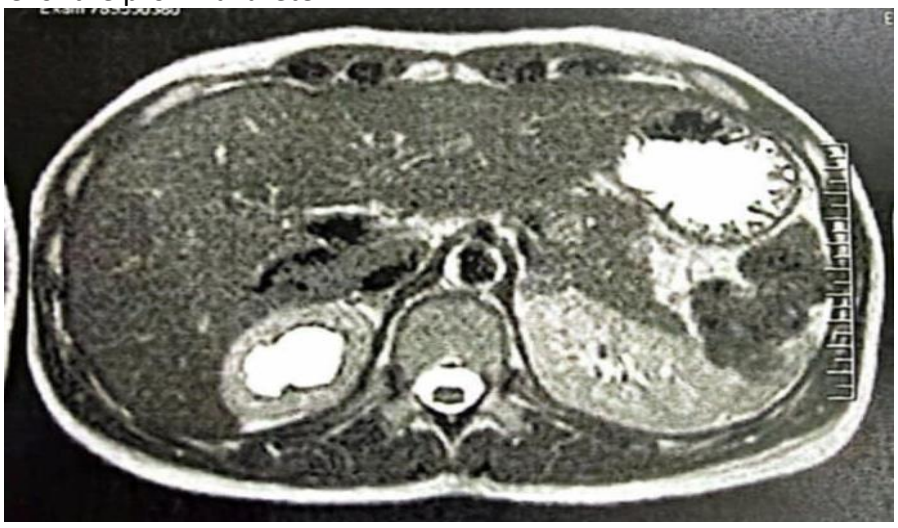

Figure 1: contrast-enhanced abdominal MRI demonstrating a nodular lesion in the pancreatic body, appearing hyperintense with mild gadolinium enhancement, without peripancreatic fat infiltration.

Endoscopic ultrasound further revealed multiple well-demarcated hypoechoic lesions in the pancreatic head, uncinate process, and tail, ranging from 5 to 9 mm, consistent with multifocal pNETs.

Fine-needle aspiration was performed, and histopathology confirmed a well-differentiated pNET (WHO Grade 1) with a low proliferation index and positive immunohistochemical staining for chromogranin A, synaptophysin, and neuron-specific enolase.

Somatostatin receptor scintigraphy demonstrated increased tracer uptake in all pancreatic lesions, without evidence of distant metastases.

Endocrine evaluation revealed a 13 × 18 mm sellar and right laterosellar mass with endosphenoidal extension, demonstrating contrast enhancement after gadolinium injection. The lesion infiltrated the right cavernous sinus, encased the intracavernous segment of the internal carotid artery, and displaced the pituitary stalk superiorly, consistent with pituitary adenoma (Figure 2).

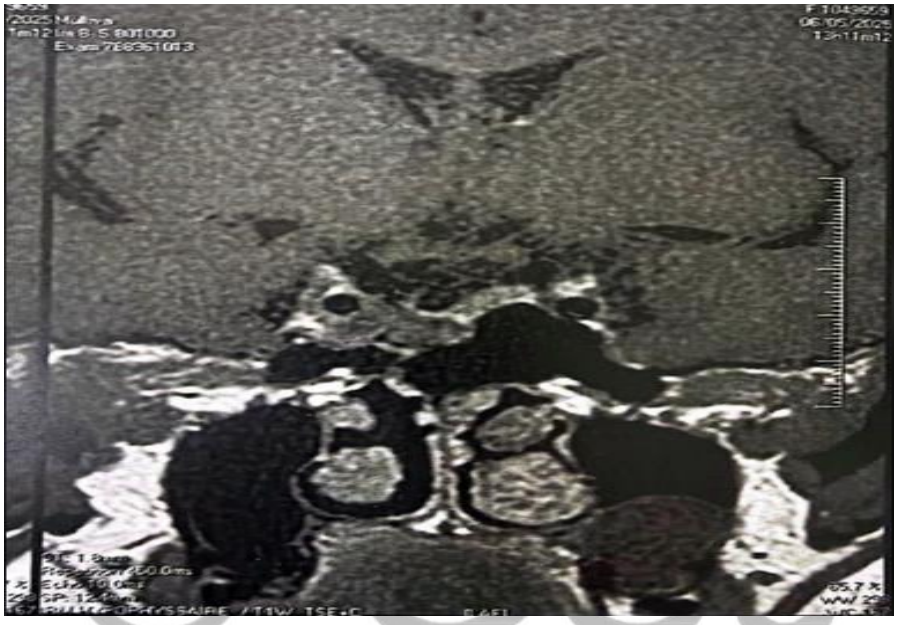

Figure 2: Contrast-enhanced pituitary MRI revealed sellar and right laterosellar mass with endosphenoidal extension, showing contrast enhancement after gadolinium administration.

Biochemical analysis also revealed hypercalcemia (11.2 mg/dL) with elevated parathyroid hormone (118 pg/mL), confirming primary hyperparathyroidism. Serum prolactin was markedly elevated (141 ng/mL). Chromogranin A was 320 ng/mL; while fasting glucose was normal at 88 mg/dL. The combination of multifocal pancreatic neuroendocrine tumors, prolactinoma, and primary hyperparathyroidism established the diagnosis of MEN1.

The patient was managed within a multidisciplinary framework involving endocrinology, oncology, surgery, radiology, and urology teams. Given the functional nature of the pancreatic neuroendocrine tumors and the presence of recurrent peptic ulcer disease, high-dose proton pump inhibitors were initiated, combined with eradication therapy for Helicobacter pylori, resulting in effective control of gastrointestinal symptoms.

In view of the diagnosis of a functional pancreatic gastrinoma, somatostatin analog therapy was introduced to control hormone secretion and stabilize tumor growth. Considering the small size (5–9 mm), multifocal distribution, and absence of metastatic disease, pancreatic surgery was deferred in accordance with current MEN1 management guidelines, favoring a conservative approach with active surveillance.

For the prolactin-secreting pituitary adenoma, dopamine agonist therapy was initiated as first-line treatment, leading to a marked biochemical response. Regarding primary hyperparathyroidism, surgical management was indicated due to persistent hypercalcemia and early renal involvement. The patient underwent subtotal parathyroidectomy, and histopathological examination confirmed parathyroid hyperplasia.

Urological evaluation was arranged for associated ureteral obstruction, and the patient benefited from specialized urological management, with no subsequent renal complications.

Outcome and Follow-up

Biochemical follow-up demonstrated significant improvement across all affected endocrine axes. Serum prolactin levels decreased to 69 ng/mL, indicating sustained control of the prolactinoma. Chromogranin A levels declined to 175 ng/mL, reflecting biochemical response of the pancreatic neuroendocrine tumors. Following parathyroid surgery, corrected serum calcium normalized to 9.6 mg/dL, and parathyroid hormone levels decreased to 54 pg/mL, confirming effective treatment of primary hyperparathyroidism.

Serial imaging with CT, MRI, and endoscopic ultrasound showed that pancreatic lesions remained small, multifocal, and radiologically stable, without evidence of locoregional progression or distant metastases. Consequently, pancreatic surgery has not been required.

After 4 years of follow-up, the patient continues under regular multidisciplinary surveillance and remains clinically stable, with good quality of life and no new MEN1-related manifestations. Genetic counseling and screening of first-degree relatives were recommended to allow early detection and appropriate management of MEN1-associated lesions.

Discussion:

This case highlights several important clinical points. MEN1 may present in a sporadic form, even in the absence of a family history, underscoring the need for a high index of suspicion. Multimodal imaging, including magnetic resonance imaging, endoscopic ultrasound, and functional imaging, is essential for the detection and characterization of small, multifocal pancreatic neuroendocrine tumors.

In the present case, the association of recurrent duodenal ulcer disease, multifocal well-differentiated pancreatic neuroendocrine tumors, a prolactin-secreting pituitary adenoma, and primary hyperparathyroidism fulfilled the clinical criteria for MEN1. Recurrent peptic ulcer disease strongly suggested a functional gastrinoma, consistent with a Zollinger–Ellison syndrome phenotype, which is commonly encountered in MEN1 and represents a major source of morbidity (3,5).

Surgical resection of the pancreas was not performed, despite the presence of a gastrinoma, because the tumors were small (5–9 mm), multifocal, and localized, and current MEN1 management guidelines recommend conservative management with medical therapy and surveillance for tumors <2 cm(1,4). Surgery is typically reserved for tumors ≥2 cm, rapidly growing lesions, or those causing uncontrolled hormone-related symptoms. The patient's gastrin levels and ulcer symptoms are currently controlled with medical therapy, including high-dose proton pump inhibitors and H. pylori eradication, supporting a non-surgical, supporting a non-surgical, surveillance-based approach.

This approach minimizes the risk of pancreatic insufficiency and other complications while providing effective control of the functional tumor. Periodic imaging and biochemical monitoring are planned to detect any tumor growth or new functional activity that might warrant surgical intervention in the future.

The demonstration of somatostatin receptor expression further supports the use of somatostatin analogs, which may contribute to tumor stabilization and symptom control (4).

Pituitary adenomas in MEN1 are most commonly prolactinomas and tend to be larger and more invasive than sporadic counterparts (1,6). In this patient, elevated prolactin levels and pituitary imaging findings were consistent with a prolactinoma. Dopamine agonist therapy was initiated as first-line treatment, in accordance with current recommendations, with surgery reserved for medically refractory cases or lesions causing mass effect.

Primary hyperparathyroidism (PHPT) is the most frequent and often earliest manifestation of MEN1 (1,2), and it may be clinically silent despite causing progressive complications. In the present case, biochemical evaluation revealed elevated calcium and parathyroid hormone levels, and imaging identified microlithiasis, confirming early renal involvement. Surgical treatment is generally recommended because of the progressive nature of the

disease and the risk of long-term complications, including nephrolithiasis progression, osteoporosis, and neurocognitive symptoms. Subtotal parathyroidectomy or total parathyroidectomy is recommended (2,6).
Genetic counseling and family screening are mandatory, even in apparently sporadic cases, as early detection of at-risk relatives allows timely surveillance and intervention.

Conclusion

This case illustrates that multidisciplinary, individualized management combining medical therapy, surgery, and surveillance can achieve effective disease control, even in patients with functional and multifocal tumors.

Acknowledgment

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