International Surgery Journal
Duraisamy V et al. Int Surg J. 2026 Sep;13(9):1773-1777
https://www.ijsurgery.com
pISSN 2349-3305 | eISSN 2349-2902
DOI: https://dx.doi.org/10.18203/2349-2902.isj20263011

Case Report

From obstructive uropathy to syndromic suspicion: a case of multiglandular primary hyperparathyroidism and pancreatic nodule mimicking multiple endocrine neoplasia type 1

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Received: 10 June 2026
Revised: 15 July 2026
Accepted: 24 July 2026

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ABSTRACT

Multiple endocrine neoplasia type 1 (MEN-1) is an autosomal dominant disorder characterized by synchronous or metachronous tumor development within the parathyroid glands, enteropancreatic neuroendocrine framework, and anterior pituitary. A 48-year-old female presented with severe bilateral loin pain and anuria lasting 1 month. Comprehensive imaging revealed bilateral hydroureteronephrosis from staghorn and renal calculi requiring urgent bilateral double-J (DJ) stenting. Subsequent endocrine evaluations demonstrated hyperparathyroidism (PTH: 191.9 pg/ml). Targeted 99mTc-Sestamibi SPECT-CT and 68Ga-Trivehexin PET-CT localized multiple hyperfunctioning parathyroid adenomas. Systemic staging via MRCP identified an additional 1.7 cm cystic neuroendocrine tumor within the uncinate process of the pancreas, structurally mimicking a classic MEN-1 phenotypic cluster. The clinical constellation of multiglandular parathyroid adenomas/hyperplasia combined with a cystic neuroendocrine tumor of the pancreatic uncinate process presents a classic phenotypic presentation mimicking MEN-1 syndrome (Wermer's syndrome).While the pituitary MRI was completely unremarkable at this stage, the coexistence of primary hyperparathyroidism and a suspicious pNET strongly mandates long-term genetic screening (MEN-1 gene mutation analysis) and close clinical surveillance. The patient's acute obstructive uropathy has been successfully managed via bilateral DJ stenting, and metabolic control has been targeted via targeted parathyroidectomy.

Keywords: Primary hyperparathyroidism, Multiple endocrine neoplasia type 1, Pancreatic neuroendocrine tumor, Multiglandular parathyroid disease, Obstructive uropathy, Staghorncalculi, Technetium-99m sestamibi SPECT-CT, 68Ga-Trivehexin PET-CT

INTRODUCTION

Primary hyperparathyroidism (PHPT) is a frequent cause of recurrent nephrolithiasis, yet its presentation as acute obstructive uropathy with bilateral hydroureteronephrosis is relatively rare. When biochemical screens reveal multiglandular parathyroid involvement in relatively young patients, a strong clinical suspicion for a hereditary tumor syndrome must be maintained. Multiple endocrine neoplasia type 1 (MEN-1), or Wermer's syndrome, classically presents with the triad of parathyroid hyperplasia, enteropancreatic neuroendocrine tumors, and anterior pituitary adenomas.[1] Several studies confirm that primary hyperparathyroidism serves as the initial manifestation in the vast majority of patients with inherited endocrine syndromes, frequently driving severe renal

calcifications before extra-parathyroid tumours display clinical features.[2] Clinically reported studies suggest that managing syndromic variants requires a highly nuanced equilibrium between aggressive oncological surveillance and targeted surgical resections to mitigate surgical morbidity.[3]

A few studies have stressed that functional molecular imaging platforms are indispensable in avoiding extensive, non-therapeutic surgeries, particularly when unexpected extra-endocrine structural anomalies are caught incidentally.[4] This case report delineates the clinical diagnostic journey, advanced functional imaging profiling, and surgical outcome of a patient presenting with obstructive uropathy who was subsequently found to display features highly mimicking MEN-1 syndrome.

CASE REPORT

A 48-year-old female presented with severe bilateral loin pain, persistent vomiting, hematuria, and a 1-month history of progressively worsening oliguria. Initial ultrasonography and computed tomography scans demonstrated bilateral hydroureteronephrosis secondary to extensive nephrolithiasis. A large staghorn calculus occupied the right pelvicalyceal system, and multiple smaller calculi were noted in the lower pole calyces of the left kidney. To treat the acute post-renal injury and protect renal function, the patient underwent urgent cystoscopy and bilateral double-J (DJ) stenting, resulting in adequate decompression with stents successfully retained in situ.

Diagnostic workup

Laboratory results revealed a state of primary hyperparathyroidism. Biochemical analysis revealed a Serum Calcium of 10.1 mg/dl, Serum Phosphorus was found to be 2.5 mg/dl, Intact PTH of 191 pg/ml (reference range: 15-65 pg/ml), Serum Albumin of 4.7 g/dl. The calculated corrected calcium was determined to be 10.8 mg/dl, 25-hydroxy vitamin D of 27 ng/ml which was found to be insufficient, alongside a serum magnesium of 3.2 mg/dl, thyroid stimulating hormone (TSH)- 2.3 µIU/ml (Euthyroid) with Free T4- 1.1 ng/dl.

The lipid profile demonstrated total cholesterol of 191 mg/dl, high density lipoprotein of 41 mg/dl and triglycerides of 191 mg/dl. Case records were analyzed on the basis of demographic data with special reference to age, sex and incidence; All patients were examined for their presentation, course of disease, various examination and investigations. These records were also analyzed for surgical intervensions and outcome.

Multimodality imaging

A 99mTc-Sestamibi parathyroid SPECT/CT scan identified a well-defined hypodense nodule with 99mTc-sestamibi uptake located posterior to the upper pole of the left lobe of the thyroid and posteroinferior to the lower pole of the right lobe of the thyroid (Figure 1). A relatively flat-appearing lesion with faint 99mTc-sestamibi uptake located posterior and medial to the upper pole of the right lobe of the thyroid. A tiny nodularity located inferior to the anterior aspect of the lower pole of the left lobe of the thyroid showing no significant Tc-sestamibi uptake.

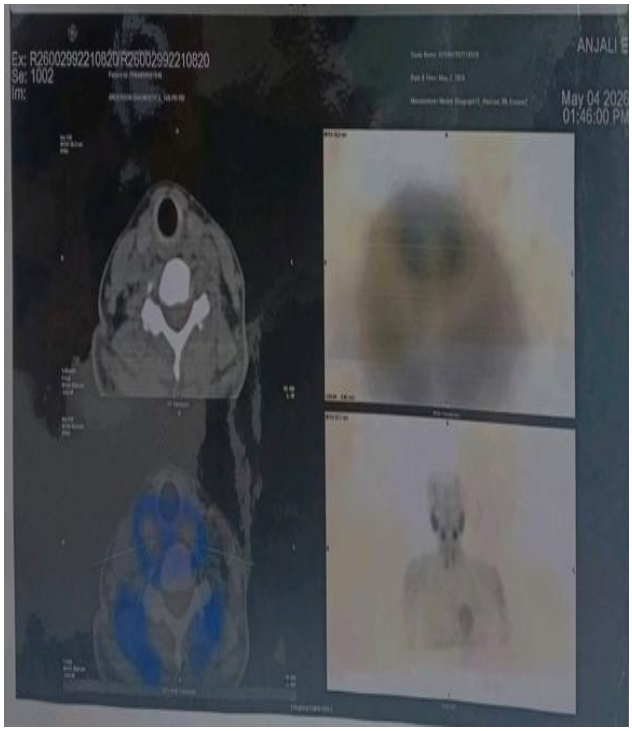

Figure 1: 99mTc-Sestamibi Parathyroid SPECT/CT scan.

MRI SELLA plain with contrast revealed Pituitary glands measuring 10.1×8.3×5.6 cm with no focal hyperintense lesion, effectively ruling out a co-existing pituitary macro- or microadenoma. Magnetic resonance cholangio-pancreatography (MRCP) revealed a well-defined T1 isointense, T2 hyperintense cystic lesion in the uncinate process of the pancreas showing diffusion restriction with low apparent diffusion coefficient (ADC) values.

A subsequent 68Ga-Trivehexin whole-body PET/CT scan demonstrated increased 68Ga-Trivehexin uptake in the well-defined hypodense nodule posteromedial to the upper pole of the left thyroid lobe, posteroinferior to the lower pole of the right thyroid lobe, and the flat lesion posteromedial to the upper pole of the right thyroid lobe-Highly suspicious for parathyroid lesions / adenomas (Figure 2).

A focal nodular appearance along the anterior aspect of the 3rd part of the duodenum / uncinate process of the pancreas with no significant 68Ga -Trivehexin avidity-Indeterminate localized pancreatic mass. A small nodularity inferior to the left thyroid lower pole with no significant tracer uptake-Indeterminate thyroid nodule.

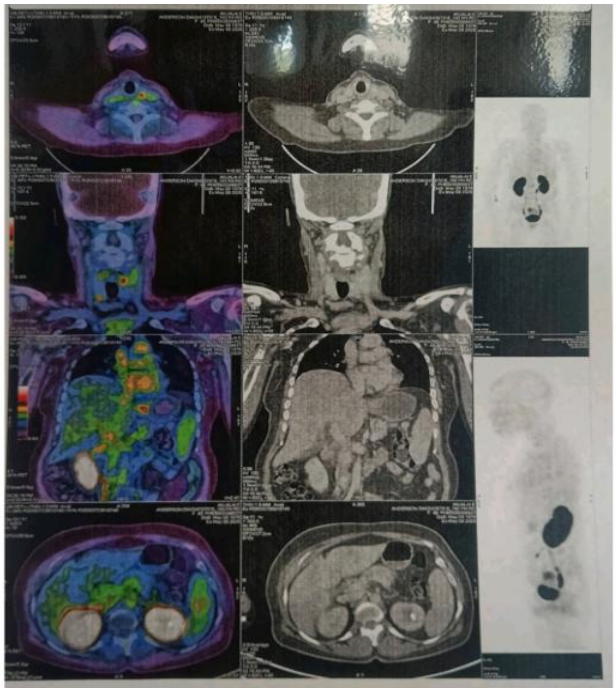

Figure 2: 68Ga-Trivehexin whole-body PET/CT scan.

Given the biochemical diagnosis of primary Hyperparathyroidism and reports of 68Ga-Trivehexin whole-body PET/CT scan, a decision was made for a comprehensive surgical exploration. The patient underwent right upper and right lower parathyroidectomy (Figure 3 and 4). Intra operative monitoring demonstrated Intact PTH at 0 hour- 93.42 pg/ml which fell significantly to 51.32 pg/ml after 20 minutes.

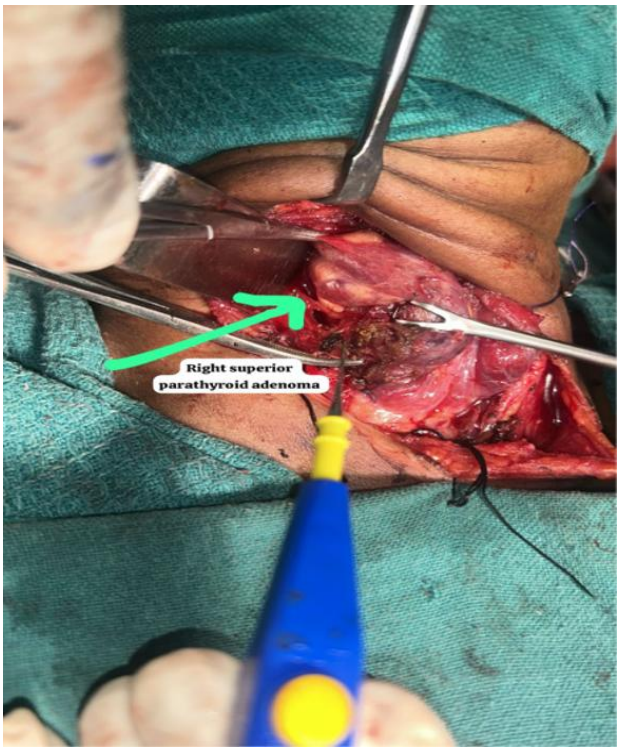

Figure 3: Right superior parathyroid adenoma.

Figure 4: Right inferior parathyroid adenoma.

Results

The surgical procedure was completed without any complications. The patient's post-operative course was uneventful. The final post-operative histopathology report (HPE) confirmed the diagnosis. The tissue from the right upper and lower parathyroidectomy was reported as Parathyroid Hyperplasia.

DISCUSSION

This clinical presentation highlights a complex diagnostic journey that initially strongly pointed toward an inherited endocrinopathy, specifically MEN-1. MEN-1 is an autosomal dominant hereditary syndrome traditionally characterized by the clinical triad of the "3 Ps": tumors or hyperplasia of the parathyroid glands, enteropancreatic pancreatic neuroendocrine tumors (pNETs), and anterior pituitary adenomas.[1] When a patient presents with synchronous tumors affecting more than one of these target organs, a germline mutation screen is strongly indicated.

In this patient's case, the initial clinical overlap was striking. She presented with primary hyperparathyroidism secondary to confirmed multi-focal, multiglandular disease (three distinct parathyroid lesions localized on her 99mTc-Sestamibi SPECT/CT and ultrasound). Simultaneously, structural abdominal imaging (MRCP) incidentally identified a 1.1×1.7×1.3 cm cystic mass within the uncinate process of the pancreas. Because this lesion exhibited restricted diffusion and low apparent diffusion coefficient (ADC) values, it carried a high radiologic suspicion for an enteropancreatic neuroendocrine tumor. Finding multi-gland parathyroid

lesions and a suspected pancreatic neuroendocrine tumor in a 48-year-old patient satisfies the clinical diagnostic criteria for MEN-1, triggering extensive systemic concern.[1]

However, cross-modality functional and molecular imaging systematically dismantled this syndromic suspicion, revealing a MEN-1 phenocopy - a situation where independent sporadic lesions mimic a genetic syndrome.[2] A dedicated contrast-enhanced MRI of the sella turcica demonstrated a completely normal pituitary architecture, ruling out co-existing adenomas. True pancreatic neuroendocrine tumors associated with MEN-1 typically feature hypervascularity and intense cell-surface marker expression.[4] To assess this, a whole-body 68Ga-Trivehexin (integrin-targeted) PET/CT scan was performed. While the scan successfully confirmed highly active, hypervascular tracer kinetics in the three suspected parathyroid glands (SUVmax ranging from 3.2 to 3.8), the 14×10 mm pancreatic uncinate lesion demonstrated no significant 68Ga-Trivehexin avidity.

The absolute lack of functional metabolic uptake strongly refutes the diagnosis of an aggressive, syndromic neuroendocrine malignancy. Instead, it indicates that the pancreatic mass is a benign, non-functioning incidentaloma - such as a simple epithelial cyst, a low-grade benign branch-duct intraductal papillary mucinous neoplasm (IPMN), or a non-secretory sporadic adenoma. Therefore, the patient represents a sporadic occurrence of multiglandular primary hyperparathyroidism coinciding with an entirely unrelated, benign pancreatic anomaly.

A systematic parameter-by-parameter comparison of our patient's findings with previous literature clarifies the clinical distinction between a true syndromic manifestation and a coincidental phenocopy. Regarding the biochemical profile, the patient's corrected serum calcium of 10.8 mg/dl and markedly elevated intact PTH of 191 pg/ml align with classic primary hyperparathyroidism, yet this degree of elevation is frequently observed in both sporadic and inherited forms. While standard guidelines for asymptomatic primary hyperparathyroidism focus on mild, progressive hypercalcemia.[3] The acute obstructive uropathy seen in our patient represents a critical, severe complication. Previous literature indicates that while nephrolithiasis occurs in nearly half of all MEN-1 patients due to long-standing parathyroid overactivity, acute anuria from an obstructing staghorn stone remains a rare presentation that demands rapid mechanical decompression prior to any metabolic intervention.

The parameter of multiglandular parathyroid involvement provides a major diagnostic pivot point. In sporadic hyperparathyroidism, a single isolated parathyroid adenoma is responsible for roughly 85% of cases, whereas multiglandular disease or diffuse hyperplasia is noted in fewer than 15% of sporadic presentations.[5] Conversely, multiglandular hyperplasia is an absolute hallmark of MEN-1, occurring in over 95% of patients by the fourth decade of life.[1] The presence of three localized parathyroid anomalies via 99mTc-Sestamibi parathyroid SPECT/CT in this 48-year-old female strongly favored a genetic syndrome. This demonstrates that structural multi-glandular disease alone cannot confirm genetic inheritance, as sporadic hyperplasia phenocopies can mirror syndromic distributions.[5]

The interpretation of the pancreatic uncinate nodule serves as the most critical parameter in differentiating these entities. In true MEN-1 syndromes, enteropancreatic neuroendocrine tumors occur in up to 70% of patients and represent a leading cause of syndrome-related mortality.[4] These syndromic pNETs are classically characterized by hypervascularity, multi-focal patterns, and an intense expression of somatostatin or cell-surface integrin receptors, making them highly avid on specialized molecular imaging like 68Ga-PET scans.[6] In our patient, the 1.7 cm cystic pancreatic mass initially raised strong diagnostic suspicion due to its restrictive diffusion patterns on MRCP. However, the complete absence of tracer avidity on the whole-body 68Ga-Trivehexin PET/CT scan strongly diverges from typical syndromic pNET findings. Historical series confirm that up to 10% of the general population may harbor entirely benign, non-functioning pancreatic incidentalomas (such as simple serous cysts or low-grade branch-duct IPMNs) that hold no clinical or genetic relationship to parathyroid disease.[4] This parameter confirms that our patient experienced a coincidental convergence of sporadic multiglandular hyperparathyroidism and a benign, non-functional pancreatic cyst, fulfilling the exact definition of a clinical MEN-1 phenocopy as described in literature.[2]

Finally, evaluating the surgical parameter confirms the clinical value of functional imaging. For true MEN-1 associated hyperparathyroidism, consensus guidelines recommend subtotal (3.5-gland) parathyroidectomy or total parathyroidectomy with autotransplantation due to an exceedingly high recurrence rate if hyperfunctioning tissue is left behind.[1] In contrast, sporadic disease allows for targeted excision of the hyperfunctioning glands identified on functional scans.[3] By employing targeted intraoperative PTH monitoring, which dropped by over 45% within 20 minutes of resection, the surgical team achieved complete metabolic resolution while avoiding the risk of permanent hypoparathyroidism. This targeted approach confirms that even when multi-organ lesions mimic an aggressive hereditary pattern, clear metabolic profiling allows for a more conservative, organ-sparing surgical strategy.

CONCLUSION

This case underscores a fundamental principle in modern endocrine oncology: structural imaging abnormalities do not equate to functional or genetic proof. While guidelines dictate an aggressive multi-organ screening protocol whenever multiglandular parathyroid disease is detected,

clinicians must look beyond structural anatomy before assigning an inherited genetic label like MEN-1.

Functional molecular imaging platforms, such as 99mTc-Sestamibi SPECT/CT and specialized targeted PET/CT scans, provide the crucial metabolic context required to differentiate true hereditary syndromes from coincidental sporadic phenocopies. Ultimately, utilizing functional metabolic imaging in this case successfully guided the surgical team toward a localized subtotal parathyroidectomy to cure her hyperparathyroidism and stop stone recurrence, while safely sparing the patient from an unnecessary, high-risk pancreatic resection.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

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Cite this article as: Duraisamy V, Lekha S, Dhanraj M, Thanislas PD, Visahini K. From obstructive uropathy to syndromic suspicion: a case of multiglandular primary hyperparathyroidism and pancreatic nodule mimicking multiple endocrine neoplasia type 1. Int Surg J 2026;13:1773-7.