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Presumed Primary Retroperitoneal Well-Differentiated Neuroendocrine Tumor with Synchronous Papillary Thyroid Carcinoma: A Case Report

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Abstract | **Case Report**

Background: Primary retroperitoneal neuroendocrine tumors are exceptionally uncommon and should be diagnosed only after careful exclusion of a more frequent metastatic or pancreaticoduodenal origin. Their association with synchronous papillary thyroid carcinoma is rarely reported and may complicate diagnostic staging and therapeutic sequencing. ***Case presentation:*** A 53-year-old man with no relevant medical history other than medically treated L4-L5 disc herniation presented with isolated left lumbar pain. Thoraco-abdomino-pelvic computed tomography revealed a retroperitoneal mass initially suspected to arise from the pancreatic uncinate process. Magnetic resonance imaging showed a retroperitoneal lesion encasing the superior mesenteric artery with associated satellite lymphadenopathy. Computed tomography-guided biopsy confirmed a well-differentiated neuroendocrine tumor, World Health Organization grade 1. Further staging with 18F-fluorodeoxyglucose positron emission tomography/computed tomography demonstrated mild metabolic activity of the retroperitoneal lesion and an incidental right thyroid hypermetabolic focus. Upper gastrointestinal endoscopy and total colonoscopy did not identify a digestive primary tumor. Cervical ultrasound classified a 9 x 7 mm right thyroid nodule as EU-TIRADS 5, and fine-needle aspiration cytology confirmed papillary thyroid carcinoma. After multidisciplinary discussion, total thyroidectomy with lymph node dissection was planned, while somatostatin receptor imaging was scheduled to complete neuroendocrine tumor characterization and guide subsequent management. ***Conclusion:*** This case emphasizes the diagnostic complexity of retroperitoneal neuroendocrine tumors and the value of comprehensive multimodal staging. Synchronous papillary thyroid carcinoma, although unusual, should be recognized because it influences treatment prioritization, follow-up strategy, and long-term oncologic surveillance.

Keywords: Neuroendocrine tumor; Retroperitoneal tumor; Papillary thyroid carcinoma; 18F-FDG PET/CT; EU-TIRADS; Synchronous malignancy; Case report.

INTRODUCTION

Neuroendocrine tumors (NETs) comprise a heterogeneous family of neoplasms arising from cells with neuroendocrine differentiation. They are most frequently encountered in the gastroenteropancreatic tract and bronchopulmonary system, whereas a truly primary retroperitoneal location is exceptionally rare [1,2]. When a NET is identified in the retroperitoneum, the main diagnostic issue is to determine whether it represents a primary tumor or a metastatic deposit from an occult gastrointestinal, pancreatic, or other primary site [2].

Because retroperitoneal NETs often grow silently, symptoms are frequently nonspecific and related to mass effect, vascular involvement, or incidental radiological detection rather than to hormonal secretion [1,2]. Histological confirmation, tumor grading, and functional imaging are therefore central to diagnosis, prognosis, and therapeutic planning [3,4].

NETs have also been associated with second primary malignancies. Several mechanisms have been proposed, including shared predisposition, treatment-related factors, field effects, and increased imaging surveillance; however, the causal interpretation remains uncertain [5,6]. Papillary thyroid carcinoma (PTC) is the most common differentiated thyroid cancer, but its synchronous discovery during the staging of a retroperitoneal NET remains unusual [7,8]. We report a case of a presumed primary retroperitoneal well-differentiated NET associated with synchronous PTC,

highlighting the diagnostic and therapeutic implications of this rare presentation.

Case Presentation

A 53-year-old male patient, with no significant medical history apart from a medically treated L4-L5 disc herniation, presented with a two-week history of isolated left lumbar pain. He reported no abdominal pain, vomiting, diarrhea, constipation, flushing, weight loss, fever, or other systemic symptoms. Physical examination was unremarkable, with stable hemodynamic and respiratory parameters. Abdominal, spinal, and general examinations did not reveal any clinically evident mass or neurological deficit.

Initial laboratory investigations were largely within normal limits, except for mild hyperglycemia, with a fasting glucose level of 1.70 g/L and glycated hemoglobin of 7%. No functional endocrine syndrome was clinically suspected at presentation.

Thoraco-abdomino-pelvic computed tomography revealed a retroperitoneal mass initially interpreted as possibly arising from the pancreatic uncinate process. Complementary magnetic resonance imaging provided a more accurate anatomical assessment and showed a retroperitoneal lesion encasing the superior mesenteric artery, associated with multiple satellite lymph nodes. This vascular relationship suggested locally advanced disease and raised the differential diagnosis of pancreatic, nodal, mesenteric, or primary retroperitoneal tumor.

A computed tomography-guided biopsy of the retroperitoneal lesion was performed. Histopathological examination, supported by immunohistochemistry, confirmed a well-differentiated neuroendocrine tumor, classified as World Health Organization grade 1 [3].

Staging was completed with 18F-fluorodeoxyglucose positron emission tomography/computed tomography (18F-FDG PET/CT). The examination demonstrated a mildly to moderately hypermetabolic retroperitoneal lesion measuring approximately 31 x 37 mm and revealed an incidental hypermetabolic focus in the right thyroid lobe. No primary gastrointestinal lesion was identified on upper gastrointestinal endoscopy or total colonoscopy.

Cervical ultrasound demonstrated a suspicious right thyroid nodule measuring 9 x 7 mm, classified as EU-TIRADS 5. Fine-needle aspiration cytology confirmed papillary thyroid carcinoma [7].

The case was reviewed by a multidisciplinary tumor board. Surgical management of the thyroid malignancy was prioritized, with planned total thyroidectomy and lymph node dissection. Additional functional imaging with somatostatin receptor scintigraphy was scheduled to define receptor expression, assess the full extent of the NET, and guide subsequent therapeutic decisions.

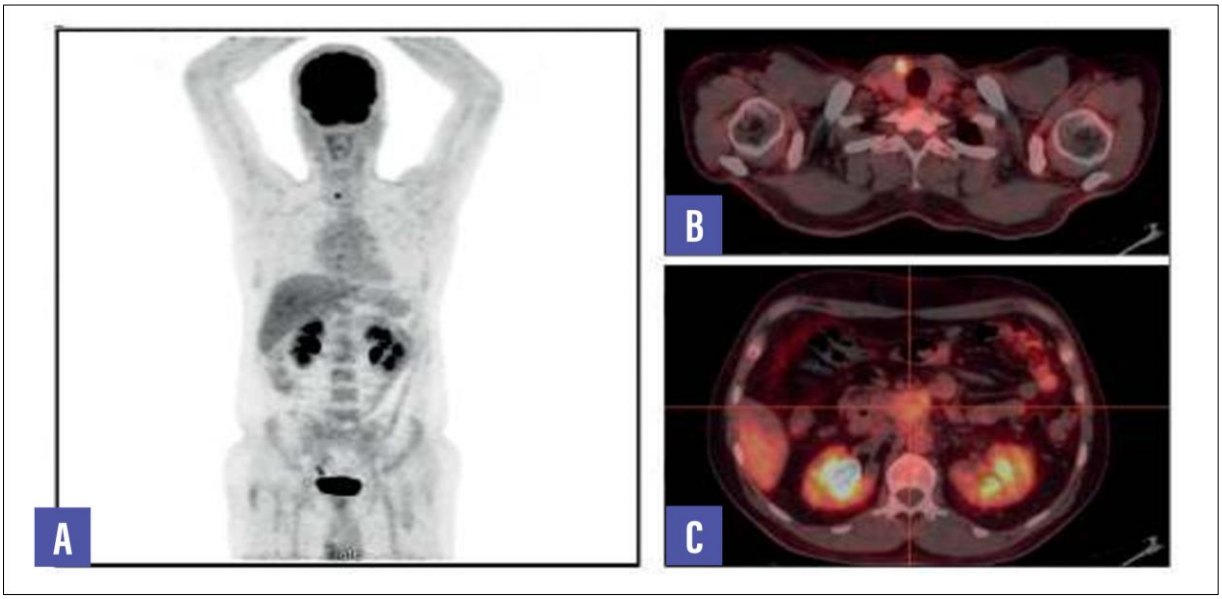

Figure 1: 18F-FDG PET/CT findings. Maximum intensity projection image showing the whole-body acquisition (A), a right thyroid hypermetabolic focus (B), and very mild hypermetabolism within the retroperitoneal lesion area (C)

Discussion

This case illustrates three clinically relevant issues: the rarity of a presumed primary retroperitoneal NET, the difficulty of excluding an occult primary digestive or pancreatic tumor, and the unexpected discovery of synchronous PTC during oncologic staging.

Primary retroperitoneal NETs are rare because the retroperitoneum is not a usual site of origin for

neuroendocrine tissue. Consequently, when a NET is detected in this region, metastatic disease from the small bowel, pancreas, stomach, colon, appendix, bronchopulmonary tract, or another occult primary tumor must be actively excluded [1,2]. In the present case, the initial CT appearance suggested a possible pancreatic uncinate process origin. MRI refined the anatomical localization by demonstrating a retroperitoneal lesion with encasement of the superior mesenteric artery and satellite lymphadenopathy. The absence of a primary tumor on upper gastrointestinal endoscopy and colonoscopy supported the hypothesis of a primary retroperitoneal origin, although this diagnosis remains one of exclusion [2,4].

Histological grading is a major prognostic determinant in NETs. A well-differentiated grade 1 tumor generally suggests slower growth and a more indolent biological behavior than higher-grade neuroendocrine neoplasms [3,4]. However, grade alone is not sufficient for therapeutic decision-making. Tumor location, size, nodal involvement, vascular encasement, symptoms, functional status, and receptor expression must also be considered. In this case, encasement of the superior mesenteric artery indicated locally advanced disease and may limit the feasibility of complete surgical resection [4].

Functional imaging is essential in the management of NETs. 18F-FDG PET/CT may be useful for staging and for detecting metabolically active lesions, but somatostatin receptor-based imaging is generally more informative for well-differentiated NETs because it assesses receptor expression and helps identify candidates for somatostatin analogues or peptide receptor radionuclide therapy [4]. For this reason, somatostatin receptor scintigraphy was scheduled after the multidisciplinary discussion.

The incidental detection of a thyroid hypermetabolic focus on 18F-FDG PET/CT was clinically important. Although many thyroid incidentalomas are benign, focal FDG uptake in the thyroid requires further evaluation because it carries a non-negligible risk of malignancy. Ultrasound risk stratification and cytological confirmation are therefore appropriate [7,8]. In this patient, the right thyroid nodule was classified as EU-TIRADS 5 and cytology confirmed PTC, leading to prioritization of thyroid surgery.

The coexistence of NET and PTC may be coincidental, particularly because differentiated thyroid carcinoma is relatively common and may be detected during extensive staging. Nevertheless, the occurrence of second primary malignancies in patients with NETs has been documented [5,6]. Proposed explanations include genetic susceptibility, shared environmental exposures, hormonal or growth factor-mediated pathways, and increased diagnostic scrutiny. In the absence of a clear hereditary syndrome or family history, this association should be interpreted cautiously, but it justifies careful long-term surveillance.

Therapeutic sequencing in synchronous tumors should be individualized. PTC is usually managed according to tumor risk, nodal status, and surgical criteria, while NET treatment depends on resectability, tumor grade, progression, symptoms, and somatostatin receptor expression [4,8]. In the present case, the decision to proceed first with thyroidectomy and lymph node dissection was reasonable because the PTC diagnosis was established cytologically and potentially curative surgery was available. Completion of NET functional imaging remained necessary before selecting observation, surgery, somatostatin analogues, locoregional therapy, or peptide receptor radionuclide therapy.

This report has limitations. The final postoperative thyroid histology, Ki-67 index details, full immunohistochemical profile, somatostatin receptor imaging results, and long-term follow-up were not yet available at the time of manuscript preparation. These data would be valuable for definitive staging, prognostic assessment, and treatment planning. Despite these limitations, the case is instructive because it demonstrates how comprehensive staging of a rare retroperitoneal NET can reveal a clinically meaningful synchronous malignancy.

CONCLUSION

Presumed primary retroperitoneal NET is a rare diagnosis that requires systematic exclusion of a metastatic or pancreaticoduodenal origin. Multimodal imaging, histopathological confirmation, and somatostatin receptor evaluation are essential for accurate staging and management. The synchronous discovery of PTC in this patient highlights the importance of investigating incidental thyroid uptake on PET/CT and reinforces the need for multidisciplinary decision-making in patients with multiple primary malignancies.

DECLARATIONS

Patient consent: Written informed consent for publication should be obtained from the patient before submission.

Ethical approval: Not applicable for a single anonymized case report according to usual institutional requirements; authors should confirm local requirements before submission.

Conflict of interest: The authors declare no conflict of interest.

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