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ENDOCRINE IMAGING

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Synchronous bilateral pheochromocytomas and bilobar medullary thyroid carcinoma revealed by ^{18}F-FDOPA PET/CT in a MEN-2A asymptomatic patient

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Type-2A multiple endocrine neoplasia (MEN-2A) syndrome is a rare autosomal dominant inheritance disease characterized by the presence of pheochromocytoma (PHEO), medullary thyroid carcinoma (MTC), and parathyroid adenoma or hyperplasia. In patients with MEN-2A, PHEO is frequently asymptomatic and bilateral, and MTC is multifocal, usually representing the first manifestation of the disease.

Herein, we present the scholar clinical case of a 38-years-old woman with MEN-2A syndrome addressed for clinical examination and primary biochemical investigation following the diagnosis of a mutation in codon 634 of gene RET exon 11. Patient showed increased plasma normetanephrine (14.7 nmol/l, normal <10.9), noradrenaline (680 ng/l, normal <400), adrenaline (257 ng/l, normal <90), dopamine (64 ng/l, normal <50), chromogranin-A (114 ug/L, normal <100), and calcitonin values (241 ng/L, normal <10). At clinical examination, small non-toxic goiter (afterwards confirmed by cervical ultrasonography) was detected. Arterial blood pressure was normal, and no Menard's clinical triad was found. Three-phases abdominal contrast-enhanced CT showed bilateral adrenal nodules of 5 × 3 cm (right) and 3 × 2 cm (left) with moderate contrast-enhancement. Adrenal lesions were characterized by intense ^{123}I-metaiodobenzylguanidine (^{123}I-MIBG) uptake. No other scintigraphic abnormality was identified, particularly in neck. 18F-fluorodihydroxyphenylalanine (^{18}F-FDOPA) positron emission tomography/computed tomography (PET/CT) completed the preoperative staging (Fig. 1), revealing intense radiotracer uptake in bilateral adrenal masses (SUVmax: 4.6 and 4.2 for right and left lesion), and in two hypodense nodules of about 1 cm of inferior pole of both thyroid lobes (SUVmax: 5.2 and 4.9). Thus, the diagnosis of synchronous bilateral PHEO and bilobar MTC was retained. Bilateral adrenalectomy was performed and pathological examination showed two tumors of 7 × 3 cm and 2 × 2 cm on right and left adrenal, without cellular atypia or capsular invasion. Final diagnosis was bilateral PHEOs with PASS score of 2. Two months later, patient underwent total thyroidectomy and bilateral lymphadenectomy. Pathological examination confirmed the presence of two nodules of right and left lobes of 1.3 × 1.1 cm and 1.3 × 0.9 cm, respectively. Cells presented with a "salt and pepper" cytoplasm, and immunohistochemistry was positive for calcitonin, chromogranin, synaptophysin and thyroid transcription factor-1 (TTF-1). Neither tumor atypia nor hyperplasia of C-cells was detected in the surrounding tissue. No lymph nodes (LN) metastasis was noted (0/28). Pathologist conclusion was bilateral MTC (pT1b(m)N0). After 5-years follow-up, patient is still in complete remission.

^{18}F-FDOPA PET/CT is an excellent imaging technique for PHEO diagnosis [1], and appears to be the most useful method for detecting MTC recurrence. At present, there is no clear advantage of PET tracers over ^{123}I-MIBG in patients with PHEO, and no definitive consensus has been reached about the added value of ^{18}F-FDOPA PET in the initial assessment of MTC. However, according to recent data, ^{18}F-FDOPA PET coupled with contrast-enhanced CT

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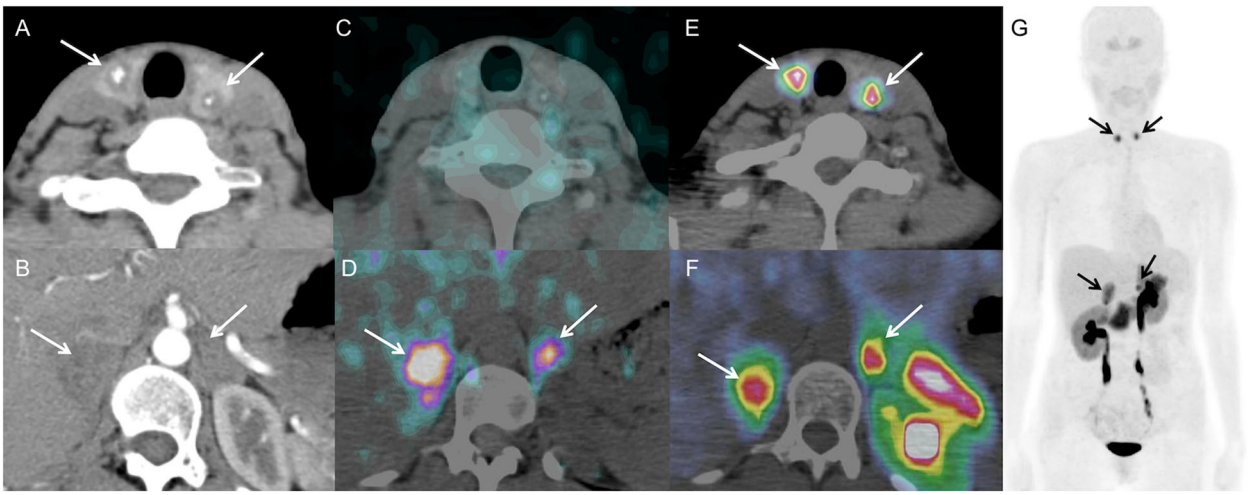

Fig. 1 Results of primary staging morpho-functional imaging in a MEN-2A patient with synchronous bilateral pheochromocytoma and bilobar medullary thyroid carcinoma (arrows). **a** Cervical unenhanced CT shows bilobar thyroid nodules with central calcification. **b** Arterial phase of contrast-enhanced abdominal CT demonstrates bilateral adrenal nodules with slight arterial enhancement. ^{123}I-MIBG SPECT/CT shows no thyroid uptake abnormalities **c**, and bilateral adrenal ^{123}I-MIBG-avid lesions **d**. ^{18}F-FDOPA PET/CT detected both thyroid **e** and adrenal tumors **f**. **g** ^{18}F-FDOPA PET maximum intensity projection (MIP)

seems more sensitive than neck ultrasonography in the detection of MTC and LN metastases at primary staging [2]. Moreover, ^{18}F-FDOPA PET/CT allows the diagnosis of distant metastases and might influences the extension of LN dissection [3]. As showed by our caricatural clinical case, ^{18}F-FDOPA PET/CT could be proposed as "one-stop shop" diagnostic investigation for primary staging in patients with MEN-2A and biochemical suspicion of synchronous PHEO and MTC. Further longitudinal investigations are necessary to exactly define the place of ^{18}F-FDOPA PET/CT in this clinical setting.

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Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

References

1. D. Taïeb, R.J. Hicks, E. Hindié, B.A. Guillet, A. Avram, P. Ghedini et al. European association of nuclear medicine practice guideline/society of nuclear medicine and molecular imaging procedure standard 2019 for radionuclide imaging of phaeochromocytoma and paraganglioma. Eur. J. Nucl. Med. Mol. Imaging **46**, 2112–2137 (2019)
2. S. Rasul, S. Hartenbach, K. Rebhan, A. Göllner, G. Karanikas, M. Mayerhoefer et al. [18F]DOPA PET/ceCT in diagnosis and staging of primary medullary thyroid carcinoma prior to surgery. Eur. J. Nucl. Med. Mol. Imaging **12**, 2159–2169 (2018)
3. L. Brammen, M.B. Niederle, P. Riss, C. Scheuba, A. Selberherr, G. Karanikas et al. Medullary thyroid carcinoma: do ultrasonography and F-DOPA-PET-CT influence the initial surgical strategy? Ann. Surg. Oncol. **25**, 3919–3927 (2018)