BILATERAL ADRENAL MYELOLIPOMAS SECONDARY TO CONGENITAL ADRENAL HYPERPLASIA: A RARE CASE OF TYPICAL ASYMMETRICAL 18F-FDG AVID

M. Kocak^{1,*}, I. Nuhoglu¹, S. Mungan², P. Duman⁴, H. Coskun¹, S. Turkyılmaz³

Karadeniz Technical University, Faculty of Medicine, ¹Department of Endocrinology and Metabolism, ²Department of Pathology, ³Department of Surgery, ⁴Kanuni Research and Training Hospital - Department of Nuclear Medicine, Trabzon, Turkey

A forty-six years old man has presented with symptoms of nausea. He has been diagnosed as congenital adrenal hyperplasia (homozygous for g.656A/C>G point mutation in CYP21A2), 3 years before admitting in a different hospital. His medical history revealed methylprednisolone use (due to lack of pharmaceutical form of hydrocortisone on Turkish market in that period) for the last year before admission. He underwent detailed medical examination and a palpable mass was noticed in his abdomen. Computed tomography scan revealed bilateral heterogeneous adrenal masses (right adrenal mass 81x42x33 mm, left adrenal mass 106x70x55 mm) containing soft tissue and adipose components and a 35 mm solid component in the right adrenal gland. Density of the solid component in the right adrenal mass was 150 HU (Fig. 1, A and B). Whole-body

18F-fluorodeoxyglucose (18F-FDG) positron emission tomography (PET) imaging revealed hypermetabolic lesion in the right adrenal gland (maximum standardized uptake value: 11.87) (Fig. 2, C and D).

Endocrinological tests assay results were as followed: ACTH: 214 pg/mL (N<46), cortisol: 5.94 µg/dL, 17-α OH progesteron: 28.6 ng/mL, (N: 0.63-2.13) dehydroepiandrosterone sulphate (DHEA-S): 29 µg/dL, (N:80-560) .

The other endocrinological workup (low-dose dexamethasone suppression test, 24-h urinary catecholamine metabolites, plasma aldosterone level and plasma renin activity) was within normal limits. He was suspected of bilateral adrenal myelolipomas secondary to congenital adrenal hyperplasia. Considering the findings (non-functional, very large mass of bilateral

Figure 1. A, Abdominal CT scan shows a 81-mm right adrenal lesion with lipoid (white arrow) and solid (black arrow) components, and a 70-mm left lipoid (B, two white arrow) adrenal lesion. CT densities of adrenal masses: for solid compartment-unenhanced CT, 150 Hounsfield units (HU); right adrenal lipoid compartment-unenhanced CT, -76 HU, left adrenal lipoid compartment-unenhanced CT, -103 HU.

*Correspondence to: Mustafa Kocak MD, Karadeniz Technical University, Faculty of Medicine, Department of Endocrinology and Metabolism, Karadeniz Technical University, Tıp Fakültesi, Endokrinoloji ve Metabolizma Hastalıkları, Bilim Dalı, Trabzon, 61080, Turkey, E-mail: mkocak19@ktu.edu.tr

Figure 2. Transaxial FDG-PET and FDG-PET/CT images and whole-body scan showed right adrenal 18F-FDG uptake (C, black arrows). Hypodense regions on CT scan were hypometabolic on FDG-PET images (D, thin arrow).

Figure 3. Biopsy shows Hematoxylin and eosin (HE) stain; E, The border between adrenal gland and neoplastic myelolipomatous area nonencapsulated tumor with mature adipose tissue and (x100). F, HE stain; Adrenal myelolipoma with lipocytes, bone marrow cells and megakaryocytes in high power field (x400).

adrenal) he was referred to general surgery and bilateral adrenalectomy was applied (1). The pathology confirmed bilateral adrenal myelolipoma (Fig. 3, E and F). He was advised to continue on methylprednisolone. His 6 and 12 month follow-ups revealed healing and event-free course.

Giant bilateral myelolipoma cases are very rare (2). The major cause is thought to be the chronic ACTH stimulation which occurs in patients with congenital adrenal hyperplasia. In our case, we think that major cause of giant adrenal mass development was lack of steroid treatment for a long period before he initiated steroid therapy and surgery seems to be effective at least for the period of follow-up (3). Due to rarity, we report a very rare case of benign unilateral hypermetabolic mass secondary to bilateral adrenal myelolipoma.

Conflict of interest

The authors declare that they have no conflict of interest concerning this article.

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