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LETTER TO THE EDITOR

Adrenal myelolipoma in association with congenital adrenal hyperplasia

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Dear Editor

The article by Decmann et al. titled "Adrenal myelolipoma: a comprehensive review" [1] was interesting and informative, in which the authors comprehensively analyzed adrenal myelolipomas by reviewing available articles on databases and their cases. Adrenal myelolipomas are the second most common primary adrenal incidentalomas following adrenocortical adenomas. According to their data, 10% of patients with myelolipoma had congenital adrenal hyperplasia (CAH) and 18.3% of all adrenal myelolipomas were associated with an endocrine disorder. Thus, they stated that the clinical picture should be considered a major factor in determining the necessity for hormonal work-up [1]. This proposal is meaningful because adrenal myelolipoma is considered a tumor that does not secrete hormones and is an exception to mandatory hormonal evaluations [2, 3]. In addition to their assertions, it is important to check whether patients with myelolipoma have CAH. This letter describes an undiagnosed man with a simple virilizing form of CAH, who experienced adrenal insufficiency after surgical resection of myelolipoma and emphasizes the importance of excluding CAH in patients with myelolipoma.

A 64-year-old man was referred to the endocrinology clinic for the evaluation of adrenal hyperplasia detected on abdominal computed tomography (CT). He was healthy with normal development during infancy and early childhood. He grew rapidly during childhood, secondary sexual characteristics appeared early, and he ceased growing at 13 years of age. He started experiencing fatigue around 30 years of age, but routine blood test results were unremarkable. Around 53 years of age, he developed dyspepsia and a decreased appetite. He was diagnosed with gastroesophageal reflux disease (GERD), but the symptoms persisted despite treatment. At 56 years of age, a left adrenal mass was discovered incidentally on abdominal CT. Laparoscopic left adrenalectomy was performed, and histopathology demonstrated myelolipoma (12.5 × 7.5 cm). Laboratory data before the surgery were as follows: 24-h urinary free cortisol, 99.8 (reference range: 75–280) μg; plasma adrenocorticotropic hormone (ACTH), 157.6 (reference range: 10–60) pg/mL; serum aldosterone, 15.5 ng/dL; and plasma rennin activity, 6.8 ng/mL/h. Abdominal magnetic resonance imaging before surgery showed a huge fatty tumor of left adrenal gland origin suggestive of myelolipoma, no visible left adrenal gland structure, and right adrenal gland hyperplasia. After the surgery, he was informed that no further investigation was required because it was a benign tumor. He experienced more fatigue and cold intolerance and was diagnosed with sensorineural hearing loss after the surgery. He received drugs for GERD occasionally, but gastrointestinal problems did not improve. Abdominal CT performed 8 years after the surgery showed diffuse nodular enlargement of the right adrenal gland and non-visualization of the left adrenal gland with multiple surgical materials in the left retroperitoneal area (Fig. 1), so he was referred for adrenal function evaluation. His height was 151 cm, weight was 54 kg, blood pressure was 105/61 mmHg, and pulse rate was 75 beats/min (regular). His blood chemistry levels were within normal ranges. His serum sodium level was 137 mEq/L and potassium level was 4.9 mEq/L. An ACTH stimulation test using 250 μg of synthetic ACTH was performed. Serum cortisol levels based on radioimmunoassays at baseline, 30 min, and 60 min were 6.97 μg/dL, 6.95 μg/dL, and 6.85 μg/dL, respectively. His baseline plasma ACTH level was 110.68 pg/mL. His baseline and ACTH-stimulated 17-hydroxyprogesterone levels were 27,500 ng/dL (reference range: 19–469) and 40,400 ng/dL, respectively. His plasma renin activity and serum aldosterone level were elevated. Genetic testing detected *CYP21A2* mutation IVS2-13A/C > G and p.I173N. IVS2-13A/C>G is one of the most frequent causes of severe CAH, and I173N is associated with a simple virilizing form of CAH.

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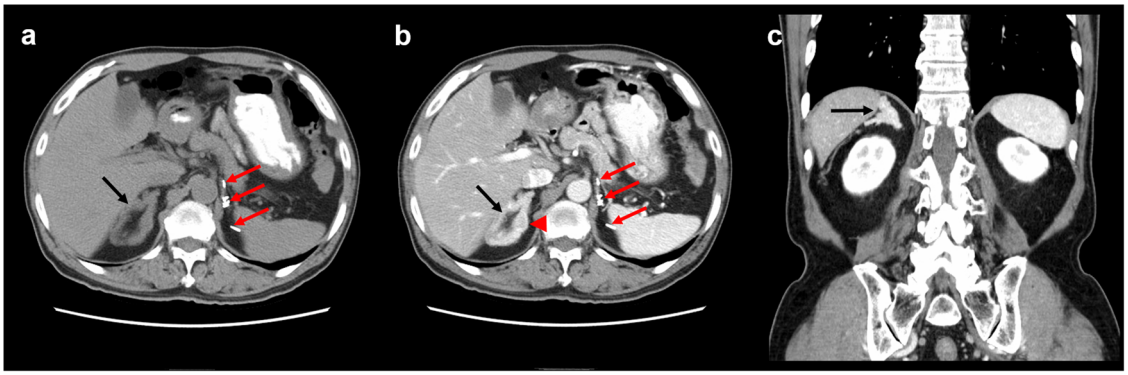

Fig. 1 Axial unenhanced **a**, axial contrast-enhanced **b**, and coronal reformatted **c** CT images of the abdomen show diffuse enlargement of the right adrenal gland (black arrows), nodular lesion (red arrowhead), and non-visualization of the left adrenal gland with multiple surgical materials (red arrows) in the left retroperitoneal area. CT computed tomography

Glucocorticoid therapy was initiated 8 years after adrenal myelolipoma resection.

A recent systemic review and meta-analysis of the relationship between adrenal incidentalomas and CAH showed that the rate (8/1000) of genetically confirmed CAH in adrenal incidentalomas is higher than the rate (1/15,000) affected by classic CAH in the Caucasian population [4]. Decmann et al. [1] showed a more detailed rate of CAH in adrenal myelolipoma. Guidelines for adrenal incidentaloma management focus on hormone excess and malignant potential [2, 3]. Recent European Society of Endocrinology and European Network for the Study of Adrenal Tumors clinical practice guidelines recommend that every patient with an adrenal incidentaloma should undergo careful assessment including clinical examinations for symptoms and signs of adrenal hormone excess [5]; however, the possible presence of adrenal insufficiency is not mentioned. The patient showed increased ACTH in the preoperative hormone test, but it seemed to exclude adrenal hormone excess. CAH and adrenal insufficiency should be excluded in patients with adrenal myelolipoma before adrenalectomy.

Compliance with ethical standards

Conflict of interest The author declares that she has no conflict of interest.

Informed consent The patient provided written informed consent for publication of this case report.

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