

1 *Clinical communication to the Editor*

2 **Adrenal and Testicular Tumor Formation due to 21-Hydroxylase**  
3 **Deficiency**

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16 **Key words:** congenital adrenal hyperplasia, hypogonadism, obesity, and  
17 glucocorticoid.

18 **Running head:** Tumors in 21-Hydroxylase Deficiency.

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22 **Authors' Contributions:** KY wrote the first draft and managed all of the  
23 submission process. HH contributed to the clinical management of the patient.

24 FO organized the manuscript.

25 **To the Editor:**

26 A 34-year-old man was referred for organic investigation of general  
27 fatigue. The patient had a medical history of 21-hydroxylase deficiency (21-  
28 OHD), for which glucocorticoid replacement therapy was commenced from birth.  
29 He was obese (154.1 cm, 71.1 kg), but results of physical examination were  
30 unremarkable. Basal plasma adrenocorticotropin (ACTH) level was elevated to  
31 183.0 pg/mL (reference range: 7.2-63.3). Serum level of basal cortisol was low  
32 (0.3 µg/dL, 7.07-19.6) and free testosterone level was also low (5.3 pg/mL), but  
33 levels of gonadotropins were normal. Computed tomography showed adrenal  
34 myelolipomas (**Figure 1A**). Magnetic resonance imaging suggested testicular  
35 adrenal rest tumors (**Figure 1B**), and semen analysis revealed oligospermia.

36 21-OHD is a common form of congenital adrenal hyperplasia in which  
37 production of cortisol and aldosterone is impaired and androgen is excessively  
38 secreted.<sup>1</sup> Insufficient glucocorticoid treatment results in continuous  
39 upregulation of ACTH secretion, leading to the development of adrenal  
40 myelolipomas and testicular adrenal rest tumors.<sup>2</sup> Men with 21-OHD have been  
41 reported to have a high risk of hypogonadism and spermatogenic abnormality.<sup>3</sup>  
42 Our case indicates the possibility of tumors developing in adult patients with 21-

43 OHD and the possibility that testicular adrenal rest tumors are associated with  
44 hypogonadism.

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46 **Ethics statement:** Written informed consent was obtained from the patient to  
47 publish this case report.

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49 **Figure Legend: Figure 1. A)** Abdominal computed tomography revealed bilateral  
50 adrenal myelolipomas (arrowheads). **B)** T2-weighted image of magnetic  
51 resonance imaging suggested bilateral testicular adrenal rest tumors.

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