A 63-year-old man with a late-onset 21-hydroxylase deficiency (21-OHD) was referred to our department due to obesity and gynecomastia (A) accompanying dyslipidemia, hyperuricemia and osteoporosis. The maturation of his genitalia and systemic pigmentation appeared in his later childhood. Glucocorticoid replacement therapy was commenced after 21-OHD had been diagnosed at 28 years of age, at which time his plasma levels of adrenocorticotropin (ACTH), cortisol and 17-hydroxyprogesterone were 108.2 pg/mL, 5.8 μg/dL and 135 ng/mL, respectively. Notably, serum testosterone was undetectable, while the serum gonadotropin levels were found to be higher than normal. MRI showed bilateral intratesticular masses with hypointensity on T2WI, thus suggesting the presence of testicular adrenal rest tumors (TARTs) (B).

Obesity associated with TART-induced hypogonadism became remarkable in his adulthood. Upregulated ACTH secretion due to insufficient treatment of 21-OHD causes ectopic adrenal cells to develop into TARTs, which compress normal tissue, thus leading to hypogonadism (1). Since late-onset hypogonadism is related to metabolic syndrome (2), clinicians should notice TART as a cause of obesity in adult 21-OHD patients.

The authors state that they have no Conflict of Interest (COI).

References
2. Katabami T, Kato H, Asahina T, et al. Serum free testosterone and...