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P22. Mister XX

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Context: Patients with Congenital Adrenal Hyperplasia due to 21 hydroxylase deficiency and whose karyotype is 46, XX are usually assigned to the female gender and reared as females. Reported here is a 46, XX patient, reared as male with male gender identity and male gender role.
Patient: A patient, 39 yeas old, 61.5 kg, 143cm , BMI 30 kg / m2. Physical examination was normal. External genitals: clitoromegaly 3.5 x 2 cm, without palpable testes. Results: Pelvical MSCT: Irregular hyperplasia of both glands (right gland diameter 78 mm, left gland diameter 119 mm). A solid formation 33x31x43 mm is found, which by its CT features resembles prepubertal uterus. The testes, ovaries, prostate and seminal vesicles were not differentiated. Laboratory analysis were normal, except for elevated triglyceride. LH, FSH concentration were supressed. ACTH and 170HP were extremely high. The concentration of testosterone was at the upper limit for healthy men. Outcome: Congenital adrenal hyperplasia was diagnosed in a completely virilised 46 XX patient. Enzymatic block and synthesis of cortisol precursors, which were further metabolized to testosterone and dihydrotestosterone led to hyperandrogenism. High concentrations of DHT led to clitoromegaly. Excess estrogen converted by aromatase from circulating adrenal androgens led to premature epiphyseal fusion.
Conclusion: Only 5% XX 46 patients with CAH throughout life develop male sexual identity and gender role such as the patient described. Also, few cases of massive adrenal incidentalomas in patients with CAH, as in our patient, are described. The contraversial nature of the issue gave no guidelines till now for medical and psychological care of 46,XX patients with CAH with male gender identity.

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