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Mayer-Rokitansky-Kuster-Hauser syndrome with hyperandrogenemia: A rare case of mullerian dysgenesis

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Mayer-Rokitansky-Kuster-Hauser (MRKH) syndrome with hyperandrogenism is a spectrum of Mullerian duct anomalies characterized by congenital aplasia of the uterus and of the upper part (2/3) of the vagina, in young women presenting infrequently with high level of testosterone. The ovaries and fallopian tubes are present. It is one of the most common causes of primary amenorrhea and affects at least 1 out of 4500 women. However only four cases were reported of MRKH syndrome associated with hyperandrogenemia in literature. The MRKH syndrome usually remains undetected up until the patient complains with primary amenorrhea regardless of normal development of secondary sexual physical characteristics. We report a case of MRKH syndrome with hyperandrogenism in a 21-year-old female who presented with primary amenorrhea, physical examination include Tanner stage 5 breasts, pubic hair stage IV with absence of cervix, short vaginal canal and no clitoromegaly. Transvaginal ultrasound revealed an infantile uterus while MRI shows small uterus with inactive endometrium and incidental findings of Tarlov cyst. Counseling, assurance and supportive psychotherapy were given to the patient. Follicle stimulating hormone, chest X-ray, 75 gram oral glucose tolerance test, BUN, creatinine, audiogram and electrocardiogram results were all within normal ranges. Chromosomal analysis was 46 XX karyotype. Serum testosterone was markedly elevated at11.1 nmol/L above the normal values for both male and female.

Biography

Melody Nethania Sutedja has completed her medical studies from Soochow University in Suzhou, China and is currently pursuing her Residency in Mariano Marcos Memorial Hospital and Medical Center in Batac, Philippines. She is currently doing Residency in Obstetrics and Gynecology.

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