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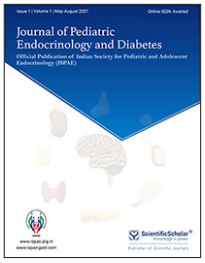

Images (Radiology/Radioisotope Scans/Fluoroscopy Images, etc.)

Testicular Adrenal Rest Tumors in a Boy with 11β-Hydroxylase Deficiency

Vijaya Sarathi

Department of Endocrinology, Vydehi Institute of Medical Sciences and Research Center, Bengaluru, Karnataka, India.

***Corresponding author:**
Vijaya Sarathi,
Department of Endocrinology,
Vydehi Institute of Medical Sciences and Research Center,
Bengaluru, Karnataka, India.

drvijayasarathi@gmail.com

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A 12-year-old boy who presented with hypertensive encephalopathy was referred to endocrinology because of hyperpigmentation. On examination, he had normal stature for age (height: 138 cm, height z-score: −1.25) but advanced bone age (18 years) suggesting severe compromise of final adult height. Tanner's sexual maturity rating was P4 and G4 (stretched phallic length: 9 cm). The right and left testes measured 5 mL and 8 mL [Figure 1], respectively. Both testes were nodular with variable consistency (hard in the upper part and soft in the lower part).

His sitting blood pressure in the right arm was 160/100 mmHg. Serum potassium was 2.67 mEq/L. Serum total testosterone was 229.77 ng/dL but the luteinizing hormone was suppressed (0.03 mIU/mL). Serum 8:00 am cortisol level was low (0.39 μg/dL) with slightly elevated serum 17α-hydroxyprogesterone (239.3 ng/dL) but markedly elevated serum corticosterone (>5000 ng/dL; normal range: 18–1970 ng/dL) and serum 11-deoxycortisol (>2000 ng/dL; normal range: 20–158 ng/dL) levels. The patient was diagnosed to have 11β-hydroxylase deficiency. Ultrasound scrotum revealed bilateral testicular adrenal rest tumors (TARTs) [Figure 1]. The patient was started on glucocorticoid replacement.

The prevalence of TART in pediatric CAH patients is around 18–24% whereas the rates up to 94% have been reported in adults.[1,2] TARTs are always benign but may cause infertility; hence, should be detected and treated early.[2] Screening for TART with testicular ultrasound should begin in adolescence and repeated every 1–2 years in asymptomatic but more frequently in symptomatic patients.[3] Optimization of glucocorticoid replacement helps to shrink early TART.[3]

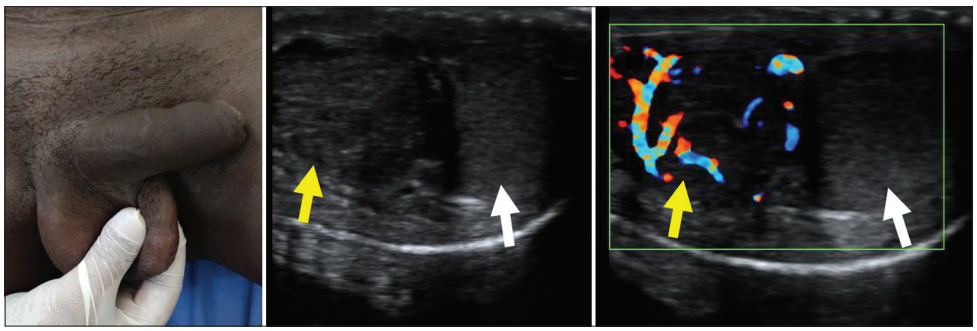

Figure 1: Clinical photograph of external genitalia (left panel) and ultrasound images showing well-defined ovoid heteroechoic lesion measuring ~ 1.5 × 1.7 cm on gray scale (middle panel) with increased vascularity on color Doppler (right panel) in upper pole of the left testis (yellow arrows) and homogenously echogenic testicular parenchyma in the lower part (white arrows).

Declaration of patient consent

Patient's consent not required as patients identity is not disclosed or compromised.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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