TESTICULAR FEMINISATION SYNDROME WITH CLITEROMAGALY

(A Case Report)

by

RENUKA SINHA
USHA SAHAY
and
CHANDRA BHUSHAN SINHA

Introduction

Testicular Feminisation Syndrome, first described by Morris in 1953 in a male pseudohermophroditism is an inherited disorder, transmitted by maternal X chromosome. It incidence rate varies from 1:20,000 to 1:64000 male births.

Case Report

Miss G.K., 19 years old female came to AMCH on 19-10-1984 for primary amenorrhoea and husky voice with hirsutism for 6 months. She was the eldest of three sisters and two brothers. No anomalies were detected in the two sisters. General examination revealed an emotionally stable, psychologically female, intelligent, tall girl with feminine atheletic body build (Fig. 1). She had downy hair over the face which was coarse over the chin. The larynx was normal. The breasts, axillary and pubic hairs were well developed. Local examination revealed well developed labia majora and minora. Left labia majora contained a tender slippery smooth oval firm lump 4 cms x 2 cms in size. It could be easily pushed up through the superficial inguinal ring into the inguinal canal of the same side. There was no impulse on coughing. In the right side of the inguinal canal just above and lateral to mons there was another small tender, firm slippery and mobile 3 x 2 cms lump. Both the lumps were tender on palpation and were found ectopic testes on clinical examination. Clitoris was enlarged about 2 cms in length. The urethal opening was feminine. The hymen was intact and circular (Fig. 2). Examination under anaesthesia revealed a cul-de-sac vagina admitting two fingers, length 6 cms. Bimanual vaginal and rectal examination revealed no cervix or uterus. The routine laboratory studies were within normal limit. The buccal smear examination for sex chromatin study was negative. 24 hours urinary 17 Ketosteroid showed a value of 17 mgms. Colour vision was normal. On 8-11-1984 under general anaesthesia bilateral orchydectomy was done and clitoris was amputated. Abdomen was opened and explored. No mullerian structures were present. Her post-operative condition was uneventful. Histopathological report of the gonads confirmed the diagnosis (Fig. 3). She was discharged with advice lynoral 1 tab. twice daily for 3 months. During follow-up there was diminition of hirsutism with slight change in hoarseness of the voice. Skin was smooth. Oestrogen was advised to be continued.

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See Figs. on Art Paper VII