

INCOMPLETE TESTICULAR FEMINIZATION

(A Case Report)

by

DEBIDAS DUTTA,* D.G.O., M.D., F.I.C.S.

and

S. B. DUTTA CHOUDHURY,** F.R.C.S.

Introduction

The term 'testicular feminization' was coined by Morris in 1953 for a hereditary syndrome characterized by individuals with testes; who present a female phenotype. A case of incomplete testicular feminization is presented for its rarity and unusual clinical interest.

CASE REPORT

A young unmarried girl, aged 20 years was admitted on 6-11-79 to the department of Obstetrics and Gynaecology. She came to hospital for primary amenorrhoea and bilateral swellings over inguinal regions. She had been noticing bilateral inguinal swellings and gradual enlargement of the phallus since the age of 14. She has two younger sisters. Menarche and growth were normal in both of them. Maternal family history was negative.

Her appearance and voice were feminine. She was 160 cm. tall, weighed 50 kg. She had slight acromegalic tendency with little prominent jaw (Fig. 1). Her skin was smooth, there was no hirsutism, scalp hair was good, axillary hair was absent and pubic hair was very scanty. The breasts were not well developed with small

nipples and poorly pigmented areolae. There were bilateral inguinal hernias, a firm ovoid mass (3.5 cm. x 2.5 cm.) was present on each side at the external inguinal ring. Examination of the external genitalia revealed enlarged clitoris (3 cm. x 1.5 cm.), well developed labia majora with less developed labia minora, vagina was 5 cm. deep (Fig. 2). On bimanual examination, cervix and uterus were found to be absent and there were no palpable adnexal masses. Other systemic examinations did not reveal any abnormality B.P. was 120/74 mm. of Hg.

Investigations: Buccal smear was negative for Bar, bodies. Drumstick pattern could not be found in peripheral smear. Vaginal smear showed predominance of intermediate cells. Intravenous pyelography revealed no abnormality. Gonadal biopsies prior to laparotomy showed testis.

Diagnosis: Incomplete testicular feminization with clitoromegaly.

Management: Both gonads were removed and herniorrhaphy was done on each side. Rudimentary epididymis and vas deferens were found with each gonad. Ovaries, tubes and uterus were found to be absent on exploratory laparotomy. Amputation of the enlarged phallus was done. She was put on oestrogen post-operatively. The development of the breasts were obvious after one month.

Histopathological examination of the gonads showed hypoplastic seminiferous tubules with absence of spermatogenesis. There was no sign of ovarian tissue (Fig. 3).

*Assistant Professor of Obstetrics and Gynaecology, Silchar Medical College, Silchar, Assam.

**Professor of Surgery, Silchar Medical College, Silchar.

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Discussion

Incomplete testicular feminization syndrome, characterized by individuals having X Y Chromosomes, two normal testicles, female external genitals and absence of the uterus and ovaries but,

unlike individuals with typical testicular feminization syndrome, they have varying degrees of mild virilization, usually clitoral enlargement and hirsutism. The case presented here meets all the criteria of incomplete testicular feminization syndrome, except hirsutism.

See Fig on Art Paper IV