A CASE REPORT OF TESTICULAR FEMINISATION

by

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Thesticular Feminisation is a rare form of intersexuality first recognised by Morris in 1953. This condition is characterised by genotype and gonadal male pattern without proper development of Wolffian structures. Urogenital sinus develops completely to a female pattern with complete absence of Mullerian structures. The person generally presents as female with primary amenorrhoea and infertility.

CASE REPORT

Mrs. A.D., 20 years old, attended the outpatients department with primary amenorrhoea. She was married for 2 years and was having normal marital relations. She was of normal built, and average health, 5′ 2½″ in height with typical physical and mental feminine characteristics. Her breasts were well developed, but rudimentary nipples, axillary and pubic hair were absent (Photograph-1). On abdominal examination, there was no in lump in hypogastrium. Both the inguinal canals contained 2 oblong, firm, smooth, mobile, non-tender swellings.

Internal genitalias showed normal vulva and clitoris, small narrow introitus. Vaginal mucous membrane was thin and vagina was represented by a blind pouch of 2″ length. Uterus and cervix could not be felt either vaginally or rectally.

The inguinal swellings were thought to be ectopic testes. With the above findings, the case looked one of the Testicular Feminisation and following investigations were done.

Pre-operative routine investigations were normal, sex chromatin pattern was negative, 17 ketosteroid — values for 24 hours urine was 19.8 mg. by Zimmermann reaction. I.P.V. was normal.

The negative chromatin pattern and raised 17 keto-steroid level confirmed our suspicion. Bilateral gonadectomy along with exploratory laparotomy was done. Both inguinal masses were removed, appeared like testis with rudimentary epididymis with gubernaculum attached to it. Vas deferens was absent on both sides, Laparotomy revealed a completely empty pelvis. The histo-pathological examination of inguinal massess confirmed the diagnosis of ectopic embryonic testes. There were testicular tubules which were lined by only Sertoli cells. Germinal cells were missing and so were the Leydig cells. (Microphotograph 2). One week post-operatively the urinary 17 keto-steroid level dropped to 16.8 mg. The patient made an uneventful recovery.

Discussion

The importance of this case lies in its rarity. Only a few cases are reported in the literature upto date. Testicular Feminisation is XY Karyotyping with negative chromatin pattern. The genotype and phenotype sex is determined by 'Y' chromosome. Absence of 'Y' chromosome causes development of female sex organs. Despite the 46 'Y' chromosomal pattern, there is failure to show the normal male development, the reasons for

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which have not been very explicitly understood.

The breast development and feminine appearance of patient is due to small amount of oestrogen secreted by adrenals and testes. If diagnosed early, the cases should be managed according to phenotype sex. The child is generally reared as a female because of phenotype female. Clitoromegaly if present, can be amputated at an early age. The accepted management is to leave the patient alone till puberty to help in proper development of breast by some of the oestrogen secreted by testes, they should then be removed because of the potential danger of their undergoing malignant changes.

See Fig. on Art Paper III