MALE HERMAPHRODITE

(A Case Report with a Review of Literature)

by

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Sex assignment and line of treatment in case of a male hermaphrodite with ambiguous external genitalia are very interesting and hence this case is reported here.

CASE REPORT

A child, aged three years, brought up as a female, was referred to the Gynaecological Endocrinology Outdoor of the K. E. M. Hospital, Bombay, for malformed external genitalia.

The child was born at full term. According to her mother, the pregnancy was uneventful and there was no history of taking any drug or exposure to radiation in that particular pregnancy, nor was there any history of bleeding per vaginam during that pregnancy.

All the mile-stones were normal, but recently she was admitted to the K. E. M. Hospital for convulsions and fever. She was diagnosed as a case of tuberculous meningitis and was being treated for the same. She is the youngest of the five siblings in her family. The four elder children are females and have no complaints.

On general examination it was revealed that this well nourished child was 31" by height and span and looked like a female child. Her respiratory, cardiovascular and gastro-intestinal systems were normal.

On gross inspection, the external genitalia resembled those of a female. The skin folds, which resembled the labia majora, were darkly pigmented in the lower part with some rugosity resembling the scrotal skin. Gonads were enclosed on either side within these skin folds. Two folds of skin resembling labia minora were also seen. On parting the labia, a rudimentary penis was seen as a penoclitoris. There was no urethral opening within the rudimentary penis. Within the two labia there was a single opening of the urogenital sinus (Figs. 1 & 4). Rectal examination did not reveal the presence of a palpable uterus in the pelvis.

On investigations it was found that the child was chromatin negative (buccal smear stained with aceto-orcein). Genitography was done by injecting a dye (Dionosil oily) through a small rubber catheter passed into the urogenital sinus as described by Shopfner (1964). As seen in the picture, there was a normal urethra and bladder, but only a small vagina. The vagina and urethra opened through a single opening into the urogenital sinus. No uterus or cervix was outlined (Fig. 2).

Gonadal biopsy was done from the gonads on both sides. The gonads were found to be testes having normal seminiferous tubules; some of them also showed spermatogenesis. The distribution of Leydig cells

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and sertoli cells was normal (Fig. 3). The biopsy of the left side accidentally contained part of the epididymis which showed normal histopathological picture.

The 17-ketosteroid estimation was 0.67 mgm./24 hours. Due to the lack of facilities it was not possible to do the detailed chromosome studies.

The case was clinically diagnosed as one of male hermaphrodite and was confirmed by the investigations mentioned above.

Discussion

The word hermaphrodite is derived from a bisexual Greek God, "Hermaphroditos", the offspring of Hermes and Aphrodite.

Male hermaphrodite can be defined as a person having the gonads of exclusively testicular tissue associated with female secondary sex organs. According to Money and many others the word pseudohermaphrodite is confusing and hermaphrodites should be classified as true, male or female, according to the histopathology of the gonads. All male hermaphrodites are chromatin negative.

A male hermaphrodite is one of the two most common types of intersexuality. The latter is further classified into two groups: (1) those with predominantly female external genitalia, and (ii) those with predominantly male external genitalia. Our patient falls in the former group. Previously, a case with testicular feminisation syndrome was also grouped with male hermaphrodites, but now-a-days it is recognised as a separate entity. Benawari (1962) recommends the classification of male hermaphrodites into three types.

According to Jones, male hermaphrodites have such ambiguous external genitalia with varying degree of development of phallus and fusion of the scroto-labial folds that there is a doubt about the sex of the individual. Rarely, they have completely normal male external genitalia with unexpected discovery of well developed Müllerian structures, usually diagnosed at the time of an inguinal hernial repair operation. Ashar and Ingle (1967) have reported such a case.

According to Overzier (1963), male hermaphrodites with predominantly female external genitalia have prominent skin folds resembling labia majora. Instead of a clitoris, there is a small phallus on parting the labia. The external orifices of the urethra and vagina may be either common as an opening of the urogenital sinus or separate. If it is separate, there is often hypospadias. The internal anatomical arrangement may be differentiated by sound, catheter or by x-ray contrast media. Laparotomy may reveal poorly developed female internal genitalia, including the uterus and the tubes. Menstruation never occurs although the Müllerian ducts may, sometimes, be well developed, but the endometrium is quite capable of responding to exogenous oestrogens.

The testes are usually intra-abdominal but in most of the cases they show some evidence of descent and are rarely situated exactly in the position of the ovaries. They may be present in the inguinal canal on one or both sides. The present case had testes in the labioscrotal folds. The testes are usually poorly developed, but in some cases well developed seminiferous tubules, spermatogonia and occasional mitotic figures may be seen. Male hermaphrodites are al-

most always sterile though few cases with uterine hernia are reported to be fertile (Overzier 1963).

At puberty the differentiation of male secondary sex characters, such as facial hair and deepening of the voice, is variable. There may be enlargement of the breasts. Vas and epididymis are present when the testes descend to the inguinal region or to the scrotum. When the testes are intra-abdominal in the position of the ovaries, the epididymis is usually absent. If it is present the vas can be traced towards the uterus between the two leaves of the broad ligament and it opens separately in the urogenital sinus.

Complications

Male hermaphrodite may have other errors of embryogenesis which may threaten life. Defects of the urinary system, bowel and anal opening, or of the heart and circulation are some of the examples.

In the post-surgical repair of an hermaphrodite as a male, complications of urinary infection may occur as a consequence of entrapment of urine in the vestigial Müllerian structures that were not recognised and excised.

Inguinal hernia is also common. The testes are liable to undergo malignant change. 50% of the neoplasms belong to the seminoma group, 20% to the teratoma and 30% are derived from variable sources (Overzier, 1963).

The most serious problems arise in an individual assigned as a boy, but without an adequate phallus.

Cases well managed from birth do not show a high incidence of neurosis or psychosis.

Aetiology of the condition is obscure. Failure of the foetal testes to produce a suppressor substance results in non-suppression of the embryonic Müllerian ducts and imperfect masculine differentiation of the reproductive tract. Foetal testicular androgen may not fail and therefore the external genitalia are not necessarily imperfect. Though occasional cases have been known in siblings the condition is usually not hereditary.

Treatment

Diagnosis of a hermaphrodite should be established as soon as possible after birth, and the sex should be assigned in such a way that there will never be any need to change such an assignment. The aim of good treatment of a hermaphrodite is to make life as normal, effective and personally rewarding as possible. Surgical, hormonal and psychological medical skills should work towards this aim.

If it is decided to rear the reported child as a female, excess of penoclitoreal tissue should be amputated and the testes removed to avoid virilisation. After puberty the individual should be treated all her life with oestrogens to develop and maintain the female secondary sex characters. The reasons for sex assignment as a female are as follows:

I. One good rule of sex assignment is — do not assign a new born hermaphrodite to the sex for which it cannot be made compatible for sexual life later on, regardless of the original genetic, gonadal and hormonal sex.

The rule applies chiefly to a male hermaphrodite with a cliteropenis who cannot be properly masculinised.

II. Corrective surgery of the genitals as a female is technically easier than as a male. An artificial vagina can be constructed successfully.

III. Serious psychiatric consequences may arise from changing the sex of rearing after the age of infancy, and instead of re-assignment of the sex the physician should exert his efforts to complete the adjustment of the individual in the sex in which the child has been reared so far. But the disaster of an assigned male with no penis is great enough to justify heroic psychological and psychiatric efforts to effect a change of psychosexual identity along with a sex re-assignment, Money (1968).

IV. As Lewis (1965) mentions, the state of the external genitalia is most important. Each case should be treated on its own merits. According to Overzier (1963), because of all the above points in these cases of male hermaphrodites the genetic and gonadal sex should be deliberately disregarded and the baby assigned as a female with eventual surgical and hormonal correction.

Psychological guidance should be given to the parents of a hermaphroditic child. Finesinger *et al* (1942), have reported about the psychiatric problems of a male hermaphroditism. When older, the affected baby will almost certainly need advice and information about his or her condition, sterility, parenthood by adoption, etc.

Summary

An interesting case of male hermaphrodite is reported with review of literature. The reasons for rearing up the individual as a female are mentioned in detail.

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See Figs. on Art Paper VI