A TRUE HERMAPHRODITE

CASE REPORT

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V.K., a Bantu 'female', aged 21, consulted her doctor to see if anything could be done about her genitalia, which at an early age she had noticed to be different from others. She was referred to Coronation Hospital.

Menstruation had commenced at 19, being a scanty loss for 2-3 days, with dysmenorrhoea, at rather irregular intervals. Intercourse had never occurred; the patient claimed to have had erections when sexually aroused by attractive females, but that more frequently she had female desires when stimulated by men.

On examination the patient was found to have the build of a lightly built male. The hair distribution was female. The breasts were fully developed as in the normal female.

The external genitalia consisted of a large phallus equivalent in size to that of a normal adult penis, lying in folds of skin corresponding to labia. A sulcus like that of hypospadias was present. Below the phallus was a small orifice through which the patient both urinated and menstruated. The anus was in the normal position (Fig. 1).

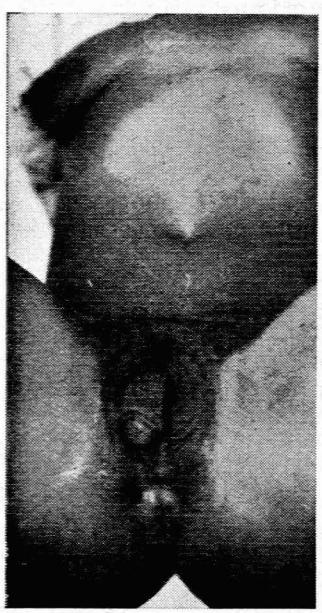

Fig. 1. External organs.

On rectal examination a small uterus was felt, with normal utero-sacral ligaments. Masses corresponding in size and position to normal ovaries were palpable. X-ray pelvimetry showed an android type of pelvis.

On 5 March 1954 a laparotomy revealed a small normally formed uterus with a small fibroid on it. The Fallopian tubes were normal.

The left gonad appeared to be a normal ovary, the right was larger than the left and was divided vertically into two portions by an obvious line of demarcation. The medial was somewhat larger than the lateral and appeared to be normal ovarian tissue, the lateral was of much softer consistency and of a greyish colour. It looked and felt like testicular tissue.

Biopsy specimens were taken from both gonads and from the skin of the phallus. They were reported upon by Dr. B. J. P. Becker, Professor of Pathology, University of Witwatersrand, as follows:

Sections of the gonad on the left side show the presence of normal ovarian tissue in which an old corpus luteum and an occasional simple follicular cyst has been observed (Fig. 2).

Sections of the gonad on the right side show the structure of an ovotestis. The *ovarian portion* of this shows the presence of a corpus luteum. The *testicular portion* shows the presence of well-developed seminiferous tubules, in which there is no evidence

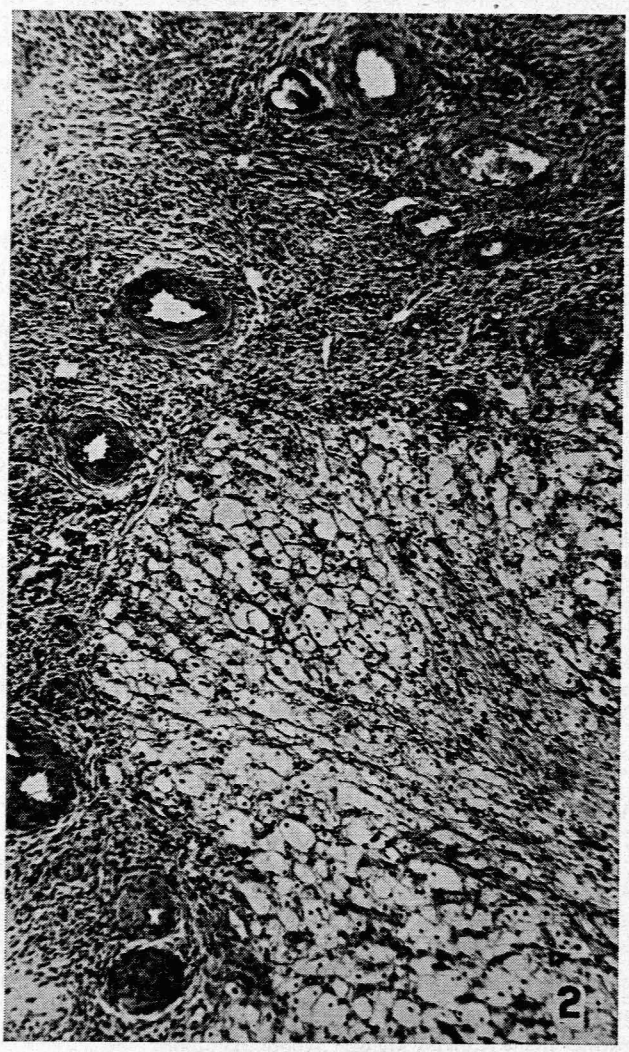

Fig. 2. Left ovary × 120. Section shows ovary and a portion of corpus luteum.

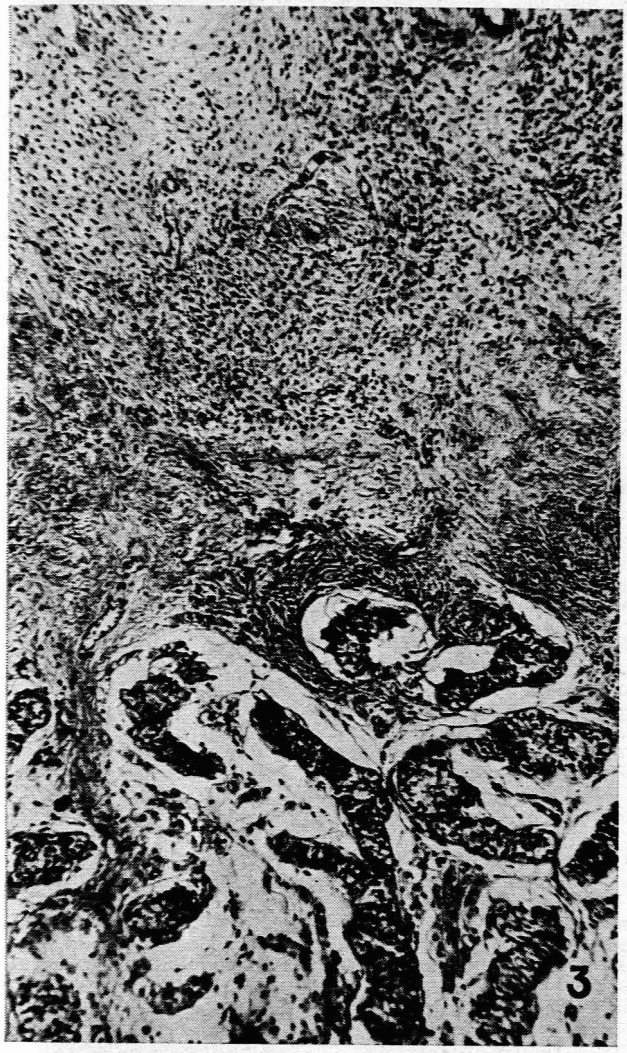

Fig. 3. Right ovotestis × 120. Section shows portions of ovarian and testicular tissue.

of spermatogenesis, and hyperplastic Leydig-cell tissue. The testicular portion is separated from the ovary by a fairly thick band of collagen. (Fig. 3.)

Sections of the skin show the structure of a *chromosomal* female.

On 26 March an operation was undertaken to modify the genitalia to the female pattern.

By means of probing it was established that the perineal orifice was a narrow vagina leading up to the cervix and that the urethra opened into it just within the entroitus. The perineum was slit posteriorly and the vagina was dilated up to the size of a No. 14 Hegar's dilator. A small nulliparous cervix was found in the vaginal vault.

The skin surface was stitched to the vaginal epithelium where the perineum had been divided, and a mould was left in the vagina for 1 week.

From the 10th day the vagina was dilated daily with glass dilators until a normal-sized vagina was obtained.

COMMENTARY

The term hermaphrodite is used to indicate that a person is bisexual in both the gonadal and secondary sex-structures. A true hermaphrodite, in the strictest sense, should be able to fertilize a female, be fertilized by a male, and fertilize itself, but no such case has ever been authoratively reported in man.

The term pseudo-hermaphrodite indicates that the person has gonads of one sex, and is bisexual in the secondary sex-structure.

True hermaphrodites can be classified as follows:

1. *Bilateral.* Testis and ovary (ovotestis) on both sides.
2. *Unilateral.* Ovotestis on one side with ovary or testis on the other side.
3. *Lateral or Alternating.* Testis on one side and ovary on the other side.

The case reported here belongs to the unilateral type with an ovotestis on the right side and a normal ovary on the left side.

The diagnosis of genetic sex of these persons cannot be made by inspection of the external genitalia. For absolute accuracy microscopic examination of the gonads is necessary. Inspection of the surface of the gonad is not sufficient and there are cases on record where mistakes have been made until thorough microscopic examination of the gonads has been carried out (Engle *et al.* 1946).

In the treatment of mature hermaphrodites and pseudo-hermaphrodites genetic sex plays a secondary role to psychologic sex (Ellis 1945, Young 1932). In this particular case the patient volunteered that her emotions were mainly those of a female, probably because of the well-developed breasts, smooth skin and facial characteristics. Because of this and also because of the development of uterus, cervix, and tubes it was decided to enlarge her vagina, to remove the large phallus, and to give her the external genital appearance of a female.

Reconstruction of the genitalia in pseudo- or true hermaphrodites should not include castration. The only indication for castration should be a pathological lesion of the gonads themselves. The development of malignant lesions in intra-abdominal testicles, although commoner than in scrotal testes, is still so rare that castration is not justified on these grounds alone. Removal of the gonads merely because they are opposed to the psychological sex will not change the emotions, but rather will take away from the patient hormonal influences that are beneficial (Brewer *et al.* 1952).

When the diagnosis of intersexualism is made at an early age it is important to establish the true sex by gonadal biopsy. This affords the opportunity of bringing up the child in its correct sex.

Detection of Chromosomal Sex in Hermaphrodites from Skin Biopsy. It has been found that the nature of the sex chromosomes (XX or XY) in an individual may be detected by examining the epidermal nuclei in a small biopsy of skin. This technique offers a new approach to the vexatious problem of hermaphrodites.

The nuclei of female specimens contain a mass of sex chromatin which is seldom seen in male specimens. The sex chromatin is believed to be derived from heterochromatic parts of the sex chromosomes. The XX

chromosomes of the female produce a chromatin mass sufficiently large to be identified, while the XY chromosomes of the male fail to produce a chromatin mass of sufficient size to be distinguished from the general particulate chromatin. (Moore *et al.* 1953, Broster *et al.* 1953).

The exact significance of chromosomal sex determination is not yet known.

The skins of the few true hermaphrodites so far examined and reported in the literature have shown the female skin structure, as in the case published here.

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