TRUE HERMAPHRODITISM

ANTONIO CARNEIRO DE MOURA AND LUDGERO PINTO BASTO

From the Division of Surgery, Department of Urology, University of Lisbon, Faculty of Medicine

True hermaphroditism, i.e. presence of both male and female gonads in the same individual, is extremely rare in the human. In Ombredanne's monograph *Les Hermaphrodites et la Chirurgie* (Paris, 1939) are collected only 19 cases proved by microscopical examination of both gonads.

Young, in his masterly monograph on genital abnormalities, was able to collect only 20 cases proved microscopically.[1, 2] In all but 2 he found an ovotestis. Since then, 17 additional cases have been reported, although not all of these were studied thoroughly enough to be regarded as instances of true hermaphroditism. Our case seems to be the thirty-ninth of the literature. McIver, Seabaugh and Mangels have reported a case which resembles our own in many respects. In all the cases reported except in our own and the one by McIver the penis was hypospadic or abnormal.

So called pseudohermaphroditism is more frequent (0.1 per cent, according to Young).

In the perusal of the literature we have made, we have been able to find only 3 cases of hermaphroditism of the lateral or alternating type (according to Krebs' classification) similar to our own (1): one by Urechia and Teposu in which there was a hypospadic penis, a testicle on the left side and a uterus ovary and tube in the right abdomen, but with no evidence of menstruation; in this case there was, also, a considerable degree of gynecomastia. Another case by Raynaud, Marill and Xicluna in which the breasts were enlarged, and a testicle was found on the left side of the size of a hazelnut, an ovary on the right side of the abdomen, the penis was hypospadic and too small to perform intercourse; in this case there was a monthly bloody discharge through the urethra. In the case by McIver *et al.* the breasts were enlarged, the penis was normal, there was 1 testicle, monthly bloody discharge through the urethra, motile sperms could be found in the ejaculated fluid and in the abdomen there was a uterus and on the right side were complete female adnexa with a pyosalpinx. In all of these three cases there was 1 ovary and 1 testicle with normal or nearly normal function. Of these only the one reported by McIver had normal external genitalia. Our own case seems to be the second one in the literature with normal external genitalia.

[1] We are indebted to Dr. Emilio de la Pena, of Madrid, Spain for his assistance in reviewing the American literature.

[2] Dr. Young in 1939 (J. Urol., v. 41, p. 781) corrected this statement, and reported that 1 case which he previously had excluded (that of Gudernatsch) should have been considered a case of true hermaphroditism, thereby making the total number of cases, up to 1937, twenty-one instead of twenty. See also article by Dr. Gudernatsch (J. Urol., **52:** 620, 1944). Editor.

CASE REPORT

J. A. T., a white young man aged 19, was examined by us at the Out Patient Department of the Division of Surgery (Prof. Reynaldo dos Santos, chief) Lisbon Faculty of Medicine. For the last 4 years he had had bloody discharges through the urethra not related to micturition. The amount of blood passed had attained at times 50 cc. These urethral bleedings were preceded by abdominal cramps on the right side and general weakness. The past few months the bloody discharges appeared periodically every 28 or 30 days and lasted about 1 week. At the age of 13 the patient noticed that his breasts were enlarging

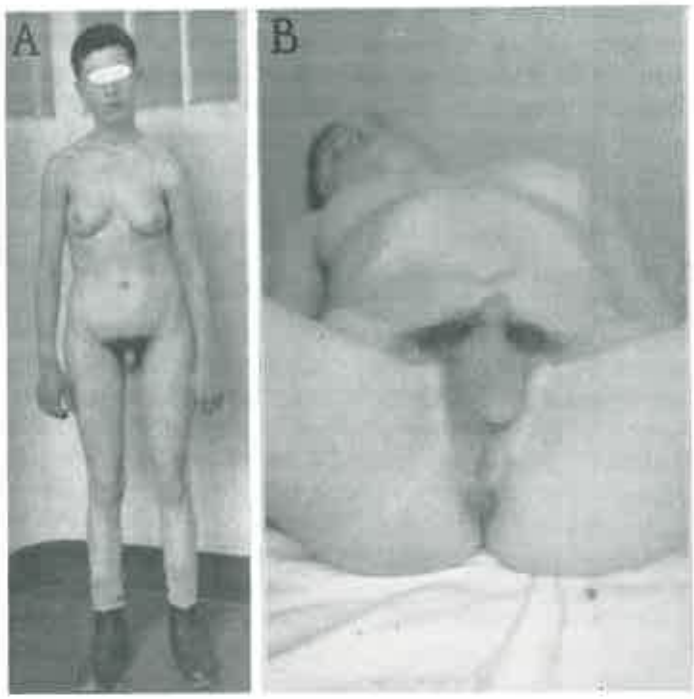

FIG. 1. *A*, Showing general development of body. *B*, External genitalia and perineum

gradually; on admission they had attained the size of those of a normal girl of his age (fig. 1). The preadolescent development was that of a normal boy, with tendency to sports. At the age of 12 his libido was manifested with erections and dreams aroused by sexual thoughts of the masculine type. At 14 he began indulging in masturbation. At 15 he noticed ejaculations for the first time; the seminal discharge was whitish and rather fluid. The patient never performed sexual intercourse because of fear of contracting venereal disease. He decided to see a doctor because his enlarged breasts caused him an inferiority complex.

The patient was one of 4 children all others of whom were normal. He had the appearance of a normal boy with a rather delicate, frail physique and broad pelvis and narrow shoulder girdle. His breasts were well developed and a female distribution of hair was present with absence of beard. His voice was

that of a normal man of his age. The patient's psychological condition seemed to be that of a normal boy although with a definite inferiority complex.

The genitalia were normal except for the absence of a gonad on the right side. On the left side a testicle and an epididymis of normal size could be palpated. At the time of his monthly discharge a tender and movable mass could be palpated in the lower right quadrant of the abdomen. Rectal examination disclosed the presence of a normal prostate with the left lobe larger than the right one (side in which the gonad is missing); the seminal vesicles and the uterus could not be palpated.

Microscopical examination of seminal fluid (Prof. Jorge Horta) showed numerous leucocytes and sperms. We were unable to carry out assays of sexual hormones. Examination of urine and blood showed no pathological changes.

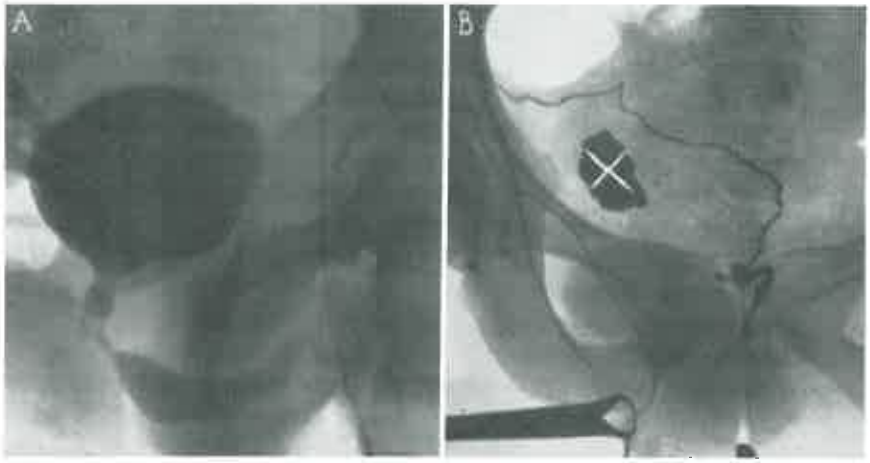

FIG. 2. *A*, Cystourethrogram obtained during menstrual period. *B*, Vaso-vesiculogram obtained by injecting iodized oil through left vas. Evidence of small seminal vesicle on right side. White cross on spot of iodized oil.

An x-ray of the skull showed no changes in the sella turcica. An intravenous urogram was also negative. An urethrogram (fig. 2, *A*) disclosed dilatation of the bulbous urethra; no communication between the urethra and the uterus could be elicited.

Cystourethroscopy showed a normal bladder. The posterior urethra was edematous which prevented visualization of openings of ejaculatory ducts or the vagina. We, therefore, were unable to attempt a vesiculography by means of catheterization of the ejaculatory ducts. Consequently we decided to obtain a vesiculogram by injecting iodized oil through a vasotomy. We took this opportunity to examine the testicle and epididymis on the left side which proved to be normal in every respect. The vesiculogram (fig. 2, *B*) showed the vas and a small seminal vesicle draining into the urethra.

A laparotomy was performed March 6, 1945. On the right side a normal uterus was found with a right ovary and tube likewise normal; absence of adnexa on the left side. The lower part of the uterus was found to proceed under

the bladder towards the posterior urethra. The uterus was amputated at the level of the bladder together with the adnexa (fig. 3). We were particularly

FIG. 3. Uterus and adnexa removed

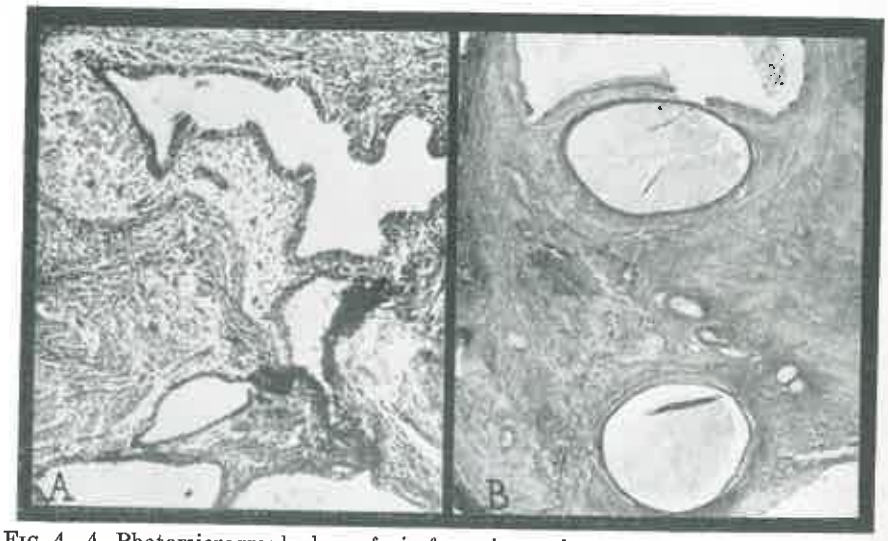

FIG. 4. *A*, Photomicrograph shows foci of ovarian endometriosis. *B*, Section of ovary with follicular cysts.

careful in closing the peritoneum and the lower end of the uterus in an attempt at preventing a possible urinary fistula, since the injection of mercurochrome through the cut end of the uterus was found to reach the bladder.

Sections of the tissue removed are shown in figures 4 and 5. The pathological studies were made by Prof. Wohlwill. The ovary was found to be active but

FIG. 5. Transverse section of uterus shows endometrium

FIG. 6. X-ray film of uterus and adnexa removed after injection of iodized oil. Tube patent.

with follicular cysts and evidence of endometriosis. The uterus showed also slight evidence of endometriosis. The cervix showed no pathological changes.

A roentgenogram of the specimen was taken after injecting iodized oil through the uterus, which showed a patent oviduct (fig. 6).

The postoperative course was uneventful. The patient resumed his sexual life by performing sexual intercourse successfully.

The breasts have shown a definite tendency to decrease in size. We suspect that it will not be necessary to resort to surgical measures to correct the gynecomastia. Urechia and Teposu as well as McIver carried out amputation of the the breasts in their cases.

COMMENT

There is no question as to our case being an example of true hermaphroditism, i.e. an individual capable of secreting simultaneously ovules and sperms, a fact that according to some writers has never been observed.

In our patient we were not dealing with a case of presence of both ovarian and testicular tissue in the same gonad (the so-called ovotestis).

The existence of a female gonad with tube and uterus was suspected by the appearance of normal and regular menstruations and proved by histological examinations. The presence of primary follicles, Graff's follicles and corpora lutea indicates that the ovary was functionally apt for reproduction. Its internal secretion is demonstrated by the existence of menstruation and of secondary sexual characters definitely feminine. On the other hand, the male gonad was of normal appearance and was attached to a normal epididymis, vas, seminal vesicle and ejaculatory duct; it was, in addition, capable of producing sperms. Moreover, there was a normal prostate and sexual tendencies of a definitely masculine type, which speaks for a normal internal secretion of the testicle.

We did not think it necessary to carry out biopsy of the testicular tissue. It is possible, however, that the gonad in the left side may be an "ovotestis". We wish to point out the fact that the follicle corresponding to the last menstruation was not found.

Unfortunately we were unable to carry out assays of sexual hormones.

As for the treatment, the libido, the normal appearance of external genitalia (male type) clearly indicated the removal of the uterus and adnexa.

SUMMARY

A case of true hermaphroditism is presented, in which the individual had a normal penis, with a testicle, epididymis, seminal vesicle and prostate and was able to secrete sperms. Besides, there was a uterus with ovary functionally active and the patient had monthly bleedings through the urethra.

This is probably the first case in which the presence of a normal ovary and a normal testicle with presence of both ovules and sperms has been observed.

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