HERMAPHRODITISMUS VERUS LATERALIS

By

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True hermaphroditism is extremely rare in the human subject. Our case is the 44th in the medical literature.

In 1937, Young, in his classic monograph, was able to collect only 21 cases with microscopic evidence, and in July 1944, Mc-Iver and his co-workers tabulated 13 additional cases. Since 1944 we have tabulated 10 more cases, including our own.

The lateral variety of this condition is even rarer: Weed and his co-workers collected in November 1947 only 11 cases of this lateral or alternating variety, with an ovary on one side and a testicle on the other. From 1947 to August 1948 we have collected 2 more cases, our present one being therefore the 14th of the lateral variety.

Our case is one of true hermaphroditism, with autopsy and microscopic examination, presenting a testicle on the right side of the scrotum and an ovary (with uterus and tube), on the left side of the pelvis.

In 44 of true hermaphroditism collected up to the present an autopsy has been performed in only 10 cases, including our own.

Hormone assays have been performed in only a few of the cases reported. In our case we assayed the gonadotrophins and the 17-ketosteroids.
The table includes the material published up to the present date.

We have no intention of publishing a general study on hermaphroditism. This subject has been well studied in Young's masterly book (1937), in the monograph by Cawadias (1946); more recently in Wilkin's book (1950), and also in several other papers. I only want to report a case with a few short comments.

**Tabulation of The Cases Previously Collected.**

<table>
<thead>
<tr>
<th>Number</th>
<th>Year and author</th>
<th>Right gonad</th>
<th>Left gonad</th>
</tr>
</thead>
<tbody>
<tr>
<td>1 to 21</td>
<td>up to 1937-Young</td>
<td></td>
<td></td>
</tr>
<tr>
<td>22 to 34</td>
<td>up to July 1944 — Mc-Iver et al.</td>
<td>Ovotestis in pelvis</td>
<td>Testis in scrotum</td>
</tr>
<tr>
<td>35</td>
<td>1944 — Moriarty</td>
<td>Ovotestis in pelvis</td>
<td>Testis in scrotum</td>
</tr>
<tr>
<td>36</td>
<td>1944 — Mc-Kenna &amp; Kiefer</td>
<td>Testis in scrotum</td>
<td>Ovary in pelvis</td>
</tr>
<tr>
<td>37</td>
<td>1944 — Mc-Kenna &amp; Kiefer</td>
<td>Ovotestis in scrotum</td>
<td>Ovary in inguinal duct</td>
</tr>
<tr>
<td>38</td>
<td>1946 — Davis &amp; Scheffey</td>
<td>Ovary in pelvis</td>
<td>Testis in a hernia</td>
</tr>
<tr>
<td>39</td>
<td>1946 — Stirling</td>
<td>Ovary in pelvis</td>
<td>Testis in pelvis</td>
</tr>
<tr>
<td>40</td>
<td>1946 — Moura &amp; Basto</td>
<td>Ovary in pelvis</td>
<td>Testis in scrotum</td>
</tr>
<tr>
<td>41</td>
<td>1947 — Weed et al.</td>
<td>Ovotestis in a hernia</td>
<td>Ovary in pelvis</td>
</tr>
<tr>
<td>42</td>
<td>1948 — Stromme</td>
<td>Testis in pelvis</td>
<td>Ovary in pelvis</td>
</tr>
<tr>
<td>43</td>
<td>1948 — Patton</td>
<td>Testis in scrotum</td>
<td>Ovary in pelvis</td>
</tr>
<tr>
<td>44</td>
<td>1951 — Berardinelli</td>
<td>Testis in scrotum</td>
<td>Ovary in pelvis</td>
</tr>
</tbody>
</table>

In 1937 Young collected only 20 cases; but in 1940 he accepted 21 cases after re-examination of the Gudernatsch case, till then considered doubtful, but already published before 1937.
CASE REPORT

J. S. O. colored, Brazilian, 49 years old, admitted to our service with a cardio-renal insufficiency.

He was of medium height, and weighed 52.4 kg. His appearance was that of a normal man for his race and age, except for the marked bilateral gynecomastia. Body hair and hair on the face was scanty; but this is quite common in his race. The pelvis was somewhat broad; the pubic hair abundant, and of a female distribution (Figs. 1 and 3).

The photograph of one of the patient’s brothers, who showed a great physical resemblance to the deceased, showed that our patient had an almost normal appearance (Fig. 2).

The patient has 3 normal brothers, none of whom has gynecomastia; he has also 4 sisters, all completely normal. One of the brothers informed us that the patient married when 23 years old, a daughter being born a year later. It was impossible to confirm this.

In addition to gynecomastia, our patient also had a supposed left "cryptorchidism", the testicle being missing on the left side of the scrotum; the right testicle was present, but was atrophied and of a soft texture.

There was no other abnormality present in the genitalia, the penis being normal in size and appearance. It was impossible to obtain sperm, because of impotence from which the patient had been suffering for some time.

His behaviour was masculine, the voice being that of a normal male, and he showed male attitudes. He was a farmer and later worked as a mason.

There was no mention of urethral bleeding, though he complained of periodic colic in the left iliac region.

Hormone Assays: — Gonadotrophins: 157 M. U. in 24 hours (ethyl alcohol precipitation, weight of the uteri of infantile mice, 72 hours test). 17-ketosteroids: 1 mg. in 24 hours expressed in androsterone (Zimmermann reaction, after Callow, Callow & Emmens).

Biopsy of the mamma: »In the middle of the connective tissue we observe some ducts lined with two or more layers of epithelium cells, presenting frequent eosinophilic secretions in the form of a projection towards the lumen. In some ducts there is hyperplasia, with formation of epithelial buds on their walls« (Fig. 9).

Biopsy of the testis: Under the thickened albuginea one is immediately impressed by the large number of interstitial cells, which actually form nodules, disseminated through out the whole organ. In the middle of this interstitial cellular mass there are found seminiferous tubes, all of them lined with Sertoli cells. There were no signs of any seminal elements. Some tubes presented a marked
hyalinization and thickening of the boundary membrane. The interstitial cells are polyhedral, and have round or kidney shaped nuclei; some of them are bi-nucleate and the cytoplasm foamy. (Biopsy Dr. G. Gouveia; pathology Dr. Daccorso). (Fig. 8).

**In summary:** — Canalicular gynecomastia. Atrophy of the seminal element and marked hyperplasia of the interstitial cells.

**Cardio-renal insufficiency and death.** — The patient entered our ward with the picture of cardio-renal insufficiency: — dyspnea on exertion, paroxysmal attacks of dyspnea at night; edema of the legs; hypostenuria; hepatomegaly; nitrogen retention (urea 105 mg. per cent; creatinine 2.8 mg. per cent; uric acid 7.3 mg. per cent; electrocardiogram: evidence of lesion of the myocardium (complete block of the right branch of the bundle of His); hypertension (Mx 250—Mn 130 mm. Hg); signs of arteriosclerosis in fundus oculi; a hypo-chromic anemia (3,300,000 per cubic mm. red cells, hemoglobin 56 per cent). He died of acute pulmonary edema.

**Autopsy and Pathology.** — (By Dr. Thales Martins, Director of the Research Department of the »Instituto de Endocrinologia«). — 2 masses were found in the left iliac region, linked to each other by a cylindrical cord, which extended to the prostatic region. It was not a patent duct, but a solid cord of connective tissue, with occasional muscular fibers. The upper structure was an ovary, containing numerous corpora albicantia (Fig. 7).

About 5 mm. below, and linked to the same cord, was the other olive shaped body; macroscopic section showed a thick wall of muscular structure, and a central vacity. In fact, after histological study, this formation was proved to be a uterus, with myometrium, endometrium and well developed glands (Fig. 5 and 6).

No seminal vesicles could be identified, but the prostate, although small and sclerosed, preserved, in several parts, groups of glandular tissue.

The adrenals weighed altogether 16.1 gm. Sections in several directions, macroscopic or after histological examinations, showed no significant changes, and no foci of hyperplasia or tumour growth (Fig. 10).

In the adenohypophysis a pronounced acidophilia was found; basophiles and chromophobes could only be found in some fields, arranged in nests or in follicles containing stainable colloid like material (Figs. 11 and 12).
Fig. 1 and 2.
The patient and one of his (normal) brothers.

Fig. 3.
External genitalia of the patient.
Fig. 4.
Scheme of the patient's genitals.

Fig. 5.
Uterus — longitudinal section "in toto".
**Fig. 6.**
Uterus — glands in the myometrium.

**Fig. 7.**
Ovary — numerous corpora albicantia.
Fig. 8.
Section of the testis. Interstitial hyperplasia with pronounced seminal lesion. Sertoli cells present.

Fig. 9.
Section of the mamma. Ducts lined with two or more cellular layers.
**Fig. 10.**
Left suprarenal (normal).

**Fig. 11.**
Adenohypophysis — pronounced acidophilia (Martins's hemat., methylblue, acid fuchs).
Fig. 42.
Adenohypophysis. — Field showing basophiles and chromophobes arranged in follicles (Martins's hemat., methylblue, acid funchs).

TABULATION OF CLINICAL PATHOLOGICAL FEATURES

1. Source of material ............ autopsy
2. Age ............................... 49
3. Reared as ...................... male
4. Emotions ......................... male
5. Bodily aspects .................. male externa genitalia; bilateral gynecomastia; pubis hair female.
6. Menses ........................... none
7. Phallus ........................... normal, not hypospadic
8. Vaginal opening ............... none
9. Uterus located .................. left side of pelvis
10. Right gonad ..................... testis in scrotum
11. Left gonad ...................... ovary left side of pelvis
12. F. S. H. ......................... high
13. 17-ketosteroids ................ low

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DISCUSSION

In view of the very serious condition of the patient and his short stay in our ward, it was not possible to repeat the hormone assays periodically.

We first diagnosed a Klinefelter's syndrome, on the evidence of the gynecomastia, high F. S. H., and the atrophy of the seminal elements without any hypo-leydigism. Later on we suspected a hermaphroditism, which was confirmed by autopsy.

It is difficult to explain the low titer of 17-ketosteroids in the urine, as the adrenal bodies were normal and there was a marked hypertrophy of the Leydig cells in the testis. However, we must remember that the patient had only one testicle, which was small and injured by the biopsy. We must also take into account the fact that during the period just before death the serious systemic diseases might have depressed the activity of the adrenals and the gonads; on the other hand, it is also possible that renal insufficiency might have hindered the excretion of the circulating hormones.

SUMMARY

The author presents a case of true hermaphroditism in a 49 years old negro with a male morphology (except for the bilateral gynecomastia); the penis was of normal size. There was only one small and soft testicle on the right side of the scrotum. Biopsy of the testis revealed atrophy of the tubules, presenting only Sertoli cells, and without any seminal elements; there was hyperplasia of the Leydig cells. Biopsy of the mamma presented a canalicular gynecomastia. The patient had a high F.S.H. excretion, and the 17-ketosteroids were low. There was no urethral bleeding, but the patient referred to periodic attacks of colic on the left side of the pelvis. The autopsy presented in the left side of the pelvis an ovary and uterus linked by a cord which extended to the prostatic region. All the glands were examined microscopically.
This is the 44th case of true hermaphroditism in the medical literature; the 14th of the lateral variety; the 10th with autopsy, and one of the few in which hormonal assays were performed.

REFERENCES

Mc-Kenna, C. M. & Kiefer, J. K.: J. Urol. 52, 464, 1944
Moriarty, J. D.: Am. J. Path. 20, 799, 1944.
Young, H. H.: Genital abnormalities and Related Adrenal Diseases.