

Psychological Aspects of the Testicular Feminization Syndrome

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Two cases of testicular feminization are reported, in sisters. One sister was hospitalized for a polymorphous neurotic reaction; the other was apparently healthy, with certain masculinoid traits in her behavior.

Certain aspects of the personality structures of intersexed individuals are discussed, with special reference to the testicular feminization syndrome and to the way in which psychosexual orientation develops in such cases.

Deception of one's self-image (which in the case of testicular feminization is less stressing than in other intersexual states) may or may not induce neurosis, depending on the over-all characteristics of the subject's personality.

PHYSIOLGIC ERRORS in the process of sex differentiation produce a variety of pathologic states grouped under the generic term "intersexual" and ranging from true hermaphroditism to degrees of male and female pseudohermaphroditism.

Such an intersexual state frequently brings about uncertain psychosexual orientation and, as corollaries, deficient determination of the individual's role and compromise of his self-image. These developments, and the concomitant feelings of inferiority and insufficiency, are the causes of neurotic structuralization of the personality of an intersexual subject. Cappon *et al.*¹ consider that only about 25% of such individuals enjoy a good mental state.

The testicular feminization syndrome, isolated by Shiller in 1910 and recently

studied in greater detail by, among others, Morriss,¹² Netter *et al.*,¹³ Molinoff and Armstrong,¹⁰ and Henrion,⁶ represents an extreme form of masculine hermaphroditism and is characterized by:

1. Male genetic sex.
2. Male gonadic sex: the testicles, immature to a greater or lesser degree, are found in an abdominal or inguinal position, or within the labia.
3. Hormonal excretion: in general, urinary androgen secretion is at the lower limit for males and the upper limit for females; urinary estrogens are at the upper limit for males and the lower limit for females.
4. A definitely female somatotype: postpuberal development of the breasts is normal, but axillary and pubic pilosity is reduced. The external genital organs are female, with a slight degree of infantilism; the vagina is comparatively small and enclosed in a cul-de-sac. Cells

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oscopy, or laparotomy, reveals the absence of uterus, oviducts, and ovaries.

These apparently "female" individuals are amenorrheic and sterile, and hormone treatment is not able to induce artificial cycles.

The disease is hereditary although it is not clear whether transmission is sex linked or autosomal and sex limited. It is believed that delayed menstruation, early menopause, and reduced axillary and pubic pilosity, in otherwise healthy gene-carrying women, are subclinical signs.

The pathogenesis of the disorder has not yet been fully elucidated: It is assumed that the fetal and adult tissues do not respond to androgens, but react normally to estrogens, and thus become completely feminized.

Approximately 200 cases of testicular feminization have been reported up to the present, although the incidence rate is felt to be much greater (approximately 1 per 62,400 male genotypes, according to Jagiello and Atwell¹).

With rare exceptions, individuals with testicular feminization (in which the endo- or exogenous androgens appear to be biologically inefficient) have a definite female orientation psychosexually. Many of them have satisfactory sexual relations and lead normal married lives; the vagina is seldom so small as not to permit intromission.

That such individuals are capable of a normal sexual impulse and satisfactory sexual activity raises doubts concerning the validity of Money's¹¹ assumption that androgens are the activating hormones of sexual desire in both sexes.

Since, except for amenorrhea, sterility, and reduced axillopubic pilosity, the testicular feminization syndrome does not involve other equivocal or stressing somatic attributes within the framework of a definitely female somatotype, and since appearances allow them to ignore their true condition, those affected have, as a rule, no doubts concerning their

identity and role. As a result, such individuals rarely develop the psychic anomalies and psychogenic reactions to which persons of other intersexual states are prone. In the available literature, only Martin and Grantham⁹ report a case involving character neurosis (dependence, instability, hysteroidism) and brooding over the sex uncertainty. That patient exhibited a variant of the testicular feminization syndrome in that certain signs of virilism existed (developed facial pilosity, deep voice, hypertrophic clitoris); several plastic interventions had been performed, one of which was complicated with a metrovaginal fistula.

Histories of the Cases

CASE 1. Our patient, A.M., 41 years old, has 2 sisters similarly affected. There was nothing of significance along the ascendant or collateral lines.

The child's development was normal and the family atmosphere stable and favorable. She unquestionably looked like a girl and was brought up as such; she had the normal interests, fancies, tastes, and activity of a little girl.

At puberty and adolescence she began to be attracted by boys and to flirt. Her menses never appeared and her breasts developed late (17-18 years), as did her axillary and pubic hair, which remained meager. These conditions gave her feelings of inferiority and embarrassment which caused her to avoid undressing in front of others. She especially avoided being examined by a doctor for fear of being asked about her menstrual cycle. At 20 she fell in love and married. Her sexual impulse was exclusively heterosexual and well developed, and sexual intercourse ended in normal orgasm. For several years her husband was slow, rather obtuse, uncommunicative, and neurotic.

The couple adopted a female child; although A.M. felt guilt toward her husband because of her sterility, her prime motivation in adopting a child was to satisfy her motherly feelings.

Since childhood and adolescence, A.M. had been noted for her punctuality, need of

order, and thoroughness, but also for a tendency to feel frustrated or neglected on certain occasions. As a young girl, and later, she demanded much affection and never seemed to get enough. With time, these traits became somewhat more accentuated.

She was exacting in the management of the house; she showed initiative and was an excellent housekeeper, but had no wish to lead a fashionable life. She was strict with her adopted child in her attempts to bring the child up to be like herself.

Since childhood she had had a swelling, interpreted as hernia, on both sides of the inguinal region. In 1952, the right side was operated on, and the parts extirpated from the inguinal region were found to be testicles. In 1962, a laparotomy was performed on the left side, and revealed the absence of uterus, ovaries, and oviducts.

Several months after the second intervention, the patient began to complain of hot flushes and of dyspareunia, which was caused by a decrease in the moisture of the vaginal cavity.

Her nervous disturbances dated back about 2 years, and had become more accentuated during the last months. They developed after surgery and menopause, and coincided with certain family misunderstandings. What seemed to weigh in the balance more than anything else were her conflicts with her adopted daughter; the girl, who had reached puberty, showed a tendency to become independent, and was no longer a "model" child.

The patient grew irritable, emotive, and anxious, and could not bear noises. She slept badly and had terrifying dreams, and complained of such psychosomatic disturbances as a sensation of burning in the head, precordial pains, diffuse numbness and itching, and respiratory difficulties. Behavioral changes also occurred: she became impulsive and choleric (followed by self-reproachfulness and fits of crying), and jealous (suspecting her husband of being in love with a young invalid). She developed tendencies to persecution interpretations, dysthymic states, repeated ideas of and attempts at suicide, dismal ideas on the vanity of all things, and self-depreciation. The patient grew unsettled and unpredictable; in the office she kept herself in hand and even felt

better subjectively, but at home the disturbances became more accentuated and reached paroxysms of either the choleric raptus type, or the suicidal behavioral or pithiatic seizure type.

She immediately liked the doctor who had begun treating her, and told her family clearly that he was the only person with whom she got on well and who understood her.

The psychologic tests revealed no organic signs, and the delay and bad performance that occurred are explicable in a functional depressive. Intellectually, her judgment was almost exclusively concrete.

In the thematic apperception test A.M. managed to build a "hero," although her stories and the clinical examination both supported an anxious, culpable person with tendencies toward perfectionism and selfishness; she could hardly have abstracted from her own personality in order to "live" the life of the "hero."

In the associative verbal test and sentence-completion test (modified according to Rotter), the depressive factor was accentuated, as was her powerful attachment to, and almost exclusive affective investment in, the family. Accompanying these factors were an accentuated ambivalence, a hypochondriac and misanthropic attitude, a tendency towards hyperconscientiousness, overevaluation of moral values according to a fairly conventional code, a feeling of unfulfillment in the satisfactions she had, a need for affection, and her present affective solitude.

CASE 2. M.G., one of A.M.'s sisters, 2 years younger, manifested no apparent psychic disturbance. Like A.M., she suffered from primary amenorrhea and had undergone 2 surgical interventions for extraction of the testicles from their inguinal position. Laparotomy revealed a small vagina and absence of uterus, oviducts and ovaries. Certain menopause disorders developed after castration, but she supported them very well.

Like her sister, M.G. unquestionably considered herself a woman. She was married and led a satisfactory erotic conjugal life. She was quite active at home and professionally, and especially active socially, with the energetic behavior of a hyperthymic personality. She had initiative, strongly developed sentiments of solidarity and fellow-

ship, and was open-hearted, direct in speech and behavior, and uncoquettish. She performed housework and manly jobs with the same pleasure. She liked "lording it" over the others in the household, and engaging them in different tasks so as to distribute the work justly. She said she felt better among men because they were "more outspoken and one could rely upon them."

The "heroine" in her thematic apperception test was an affectively traumatized woman: seduced, despised, tormented, shy, making mistakes easily, incomplete and unsatisfied in her home life, and sentimental and apprehensive. A slight misanthropic tinge could be discerned, counterbalanced by high moral aspirations and an intense desire to reach perfection.

Her answers to the verbal associative test and sentence-completion test were definitely optimistic in character.

The somatic examination established that each sister was unquestionably of feminine aspect, with slightly hypotrophic vulva, normal vagina, impalpable uterus, well-developed (but not hypertrophic) breasts, reduced axillary and pubic pilosity, and slight distal macroscelia. Neither sister exhibited deuteranomaly. Smears of the buccal mucosa showed Barr's bodies to be absent.

The cytovaginal smear of Patient A.M. was of the menopause type: 17-ketosteroids within normal limits (8.4 mg. according to the Dreckter method).

Comments

Hampson *et al.*,⁵ in studying the determinism of psychosexual orientation in intersexual individuals, concluded that the educational factor (i.e., the "imprinting" process that begin at birth and continues for 2 or 2½ years) is of major and almost exclusive importance, since it almost definitely establishes an individual's image of himself and of his role in life. But, irrespective of his role, which links him to a certain system of values and (to a certain extent) to conventional behavior, an individual may manifest certain psychological traits (inclinations, preferences, wishes, attitudes, way of being) of the other sex.

Some cases show, however, that the sexual role is not exclusively determined by education, and that certain discrepancies exist between this role and certain characteristics of the psychological profile. Certain cases of testicular feminization may be given as examples, just because these individuals do not comply with the general rule according to which they are unequivocally women psychosexually and from the viewpoint of psychological physiognomy.

Court-Brown *et al.*² mention, without further details, that 1 of the 6 cases they studied had the psychological profile of a male.

Koenig⁶ describes 2 sisters, both brought up as girls. The older, at the age of 13, had boyish tastes and none of the interests of girls of that age. In the Rorschach and thematic apperception tests, she adhered to the man's role, rivaling with her father and other males for female sexual objects. Her younger sister, 4 years old, had the female psychology of her age.

Decourt³ relates the case of a young girl who, although she had affirmative female aspirations (wishing to marry, to have her own children and home), was not actually attracted to boys, and had boyish interests (machinery, electricity, sports). She never showed interest in, or practiced, any of the specific pastimes of girls, or did housework. She was not coquettish. Her one girl friend, of whom she was very fond, also had certain androgynous psychologic traits.

Masculinoid psychical traits appeared in the manifest behavior, of M.G., the second of our sisters, but they may be assumed to represent the effect of a process of compensation for the deep affliction and dissatisfaction caused by the awareness of her infirm femininity. The projective tests appear to suggest this possibility.

There are cases which indicate that in the testicular feminization syndrome the impulse towards a feminine development

is strong enough to overcome the educational factors. Thellier *et al.*¹⁵ describe such an individual. This person was declared a girl at birth, but during an operation for inguinal hernia, at the age of 45 days, the true sex was detected and the child was thereafter brought up as a boy. Four plastic operations were performed up to the age of 14 in order to masculinize him, but at 19 he still had the female psychic and somatic habitus.

Conclusions

In the first part of our paper we explained why, among intersexual individuals, those with a testicular feminization syndrome are less likely to manifest personality and neurosis disturbances. It may be asserted that an infirmity is the more stressing the more apparent it is; people generally react less intensely, even to severe defects, if they can be dissimulated, and more intensely if these defects appear to disfigure their apparent image. Generally, an individual is less interested in his image of himself than in the evaluation of his image by others. This generalization also holds true for cases of Turner's syndrome: the patient wishes to follow treatment not to bring about menstruation and be able to function efficiently as women, but in order to grow; what most disturbs them is their small stature, which others can see.¹⁴

The reaction to feelings of inferiority varies from one individual to another. The presence of highly narcissistic demands may explain why certain irritating and compromising defects (such as amenorrhea, sterility, and reduced pilosity) cause neurotic dissatisfaction in some persons and not in others.

Easson⁴ showed that a doubtful or annoying self-image may bring about a weakening of the whole personality that renders the individual more sensitive to other stressing factors.

A.M.'s feeling of inferiority (and

shame because of her inferiority), accompanied by almost phobic avoidance behavior, had existed for a long time. Subsequent events experienced under the conditions of her neurotic potential brought about a polymorphous decompensation that took a depressive and hysteroid form.

Hampson *et al.*⁵ showed the necessity of applying psychoprophylactic measures in order to help develop the psychism of intersexual persons in as balanced a course as possible.

They also stressed the nocuous psychologic consequences of trying to make an individual's apparent sex conform to his genetic and biologic sex after the age at which his role and orientation have been established.

In the testicular feminization syndrome, the majority of persons affected unquestionably behave, for the most part, as women, and are so accepted by those around them with no reservations; to change their sex would be both useless and dangerous, since, because of their conformation, no intervention could make them efficiently male.

In our opinion, to tell these patients that they are men from the genetic-gonadic point of view is a mistake (made in both our cases), as the only result of this medical sincerity is that the subject is confronted with a painful and distressing reality. A delicate problem is raised, however, by the rare individual whose psychological profile is masculine.

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