Sexual delusion in a case of vaginal aplasia after surgical operation for neovagina

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Summary

The Mayer-Rokitansky-Kuster-Hauser (MRKH) is a syndrome of unknown etiology characterized by congenital aplasia of the uterus and the upper part (2/3) of the vagina in women showing normal development of secondary sexual characteristics. We report the case of a patient with vaginal aplasia and schizophrenia presenting with sexual delusion. To the authors' knowledge this is the first case to provide evidence of coexistence between MRKH and sexual delusion in a schizophrenic patient. The core of the patient's delirium was that she was having sexual intercourse with an eminent person through the big toe of her right foot. We approached this case using a neurological and a psychodynamic hypothesis. The neurological hypothesis suggests that the "deactivation" of the patient's genitalia led to an expansion of the adjacent big toe cortical area. The psychodynamic hypothesis supports that the sexual function and pleasure was partially expelled from the body image and was stored in a non sexual part of the body (i.e., big toe). Clinicians should be aware of this association and offer patients with MRKH psychological or/and psychiatric evaluation.

Key words: Mayer-Rokitansky-Kuster-Hauser syndrome; Narcissistic trauma; Neovagina; Sexual delusion; Schizophrenia.

Introduction

The Mayer-Rokitansky-Kuster-Hauser (MRKH) syndrome is a rare entity (incidence of 1 in 4,000-5,000 female births) characterized by vaginal aplasia and absent uterus or an extremely rudimentary one. Its etiology is unclear [1, 2]. In embryological terms, it is an aplasia or dysplasia of the Müllerian (paramesonephric) ducts. The external genitalia and vestibule, deriving from the urogenital sinus, are normal. The sex chromatin pattern is female, and the endocrine system is not affected. Ovarian function is normal, so that secondary sex characteristics appear on schedule. Although MRKH syndrome can be diagnosed after birth, in the majority of cases, diagnosis is made during puberty, due to primary amenorrhea or inability to complete the sexual act. Sterility also constitutes a major problem in the syndrome [3].

In one study the existence of narcissistic trauma was characteristic in all patients with MRKH syndrome [4]. It is difficult to assess the emotional trauma at diagnosis and its repercussions on these women's future life [3]. The absence of a vagina and uterus creates a feeling of dissimilarity and doubt about femininity and gender [2]. Patients often refer to the fact that they feel incomplete, present low self-esteem and express fear and concern about their sexual life and their relation to the opposite sex. Patients may show emotional instability and isolation, depression, anxiety and rarely suicidal behavior [2, 5-7].

Treatment involves the development of an operational neovagina [8, 9]. The emotional maturity of the patient seems to be more important than the type of intervention chosen (surgical or not) for the creation of a neovagina [7]. A rehabilitation process does not necessarily take place immediately after the diagnosis. Nevertheless, it has been suggested that after the patients have been informed of their condition, psychiatric evaluation followed by psychiatric consultation and psychotherapy is necessary [10]. The sooner the psychological support is provided to the patient, the better the therapeutic results become, both during and after the surgical intervention [4, 11, 12].

The aim of this case report is to present a case report of a schizophrenic patient with MPKH and to discuss the possible connections between sexual delirium and MPKH in a schizophrenic patient. With the incidence of the MRKH syndrome being 1/4,000 in female births while that of schizophrenia being approximately 1/100, the possibility of both disorders co-occurrence is exceptionally rare (1/400,000). To our knowledge, our case is the first of a simple co-occurrence between schizophrenia and MRKH syndrome.

Case Report

We present the case of a 28-year-old woman (NN) with MRKH which was diagnosed at the age of 17 during the evaluation of primary amenorrhea. After surgical creation of a neovagina following Creatsas vaginoplasty she engaged in a sexual life.

At the age of 20, while a university student, she presented with psychotic symptoms: delusions of persecution, grandeur, reference, passive control, mind reading, and thought broad casting. She believed that she was being trapped by her employers, her brain was opened by electromagnetic waves, her thoughts were withdrawn (thought withdrawal) and broad-

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casted. She thought she would become a diplomat and relate to very important political persons. Initial treatment with haloperidol and biperidine failed due to poor compliance. For the next few years, she suffered almost continuously from psychotic symptoms both positive and negative, never being able to return to her studies and subsequently to work. She was hospitalized in a psychiatric clinic (age 25). Since then she has been continuously under psychiatric observation and medication (risperidone, olanzapine, amisulpride, quetiapine and for some time antidepressants, citalopram, venlafaxine, fluoxetine) never resulting in a total remission of symptoms.

In January 2009 she was referred to our psychiatric clinic. She agreed to hospitalization, and she was cooperative. She had some negative-oriented symptoms including a slightly blunted affect, emotionally withdrawn, socially withdrawn; but mainly positive-oriented symptoms: sexual delusions of persecution, reference, thought withdrawal, tactile/kinesthetic hallucinations. One of her delirious experiences was that she had communication with Mr K (an ex-president of the USA) who cared about her and used her as a secret secretary. She believed that Mr K had caused a burning inside her brain. He communicated with her and they made love through the big toe of her right foot. The intercourse with Mr K via the patient's lower limb represented the core of her delirium, an event about which she spoke exclusively to her psychiatrist. The patient mentioned that often when she was lying down, she was thinking of Mr K and she was fantasizing about making love with him. Specifically, she mentioned that she felt an orgasm by stimulation of the big toe of the lower limb through which she was having sexual intercourse with him.

She was diagnosed with schizophrenia, paranoid type, treatment resistant and she was started on the atypical antipsychotic clozapine. During her hospitalization psychosocial interventions occurred including individual psychotherapy, group psychotherapy and expressive group therapy. Positive symptoms were partially limited and negative symptoms had ameliorated.

Unfortunately, NN denied any further examinations, such as for example somatosensory evoked potentials from the corresponding areas which might have been relevant to details of her delirium.

Discussion

A literature review revealed limited evidence on the psychological impact of MRKH diagnosis [12]. Symptoms of anxiety and depression observed in patients with MRKH are more severe than those in the general population and less severe than those in the psychiatric population [6]. Informing the patient about the diagnosis of the syndrome causes narcissistic trauma, disrupts the self image and leads to a deep feeling of incompletion. Feelings of insufficiency/deficiency may follow the patient for years due to their sterility [6, 13].

In this case, psychosis followed MRKH and the surgical creation of a neovagina. It is known that a severe corporeal deficit, illness, or amputation may either precede or/and provoke a psychosis. However this putative cause-effect relation is often obscure and vague. In this case, the putative cause (aplasia and then neovagina) is focused on an extremely crucial area-organ for a woman's sexual life and identity maturation and the phenomenology of the assumed effect (i.e., the psychosis) is dominated by a sexual delirium.

An interesting neurological approach to this case is provided by the remapping, reorganization hypothesis based on the plasticity of the central nervous system (CNS), a hypothesis developed by Merzenich *et al*. [14]. In the somatosensory cortex and according to the Penfield homunculus, the genitals (vagina and penis) are adjacent to the foot and especially to the big toe [15]. It is well known that extensive training of the thump leads to an enlargement of the corresponding cortical area [14]. The plasticity of the CNS and the reorganization of the cortical representation areas has already been described as the main mechanism for explaining the phantom lower limb when some points in the genitalia are stimulated, in a word "from the phantom leg to the adjacent genitalia" [15]. One wonders whether the "deactivation" of NN's genitalia led to an expansion of the adjacent big toe area. Consequently, the patient's delirious replacement is different from that observed by Aglioti *et al*., i.e., from the phantom vagina to the adjacent big toe. Furthermore, this putative remapping effect appears two to three years after the neovagina operation and only during delirium. NN never reported any kind of vagina and big toe interplay.

In parallel to the neurological hypothesis, a psychoanalytically based hypothesis can be built with regard to the correlation between vaginal aplasia and its surgical correction, and consequent changes in sexual life and sexual delusion. Psychoanalysts working with psychotic adolescents describe that changes of the body image will occasionally lead to threatening the person's ego cohesion. Laufer and Laufer [16] and Laufer [17] have suggested that overwhelming anxiety is produced from the new physical experiences of the sexual body and the inability, in some cases, to be gradually integrated. The so-called "idealized prepubertal body image" is attacked from within. The ego is forced to defend itself against external reality and the person's actual body to maintain the fantasy of the idealized body image. This "intrusion" leads to a psychotic core.

In our case the surgical correction led to a violent - for NN's ego – transformation of body image to sexual body. We suggest that during the preceding period in adolescence, a non-sexual, ideal body image was maintained, which may have allowed NN to feel in unity with the ideal mother. This omnipotence served as an inner vulnerability, so that changes in the body and in the feelings could not be integrated in the internal body image. Therefore, the sexual function and pleasure were partially expelled from the body image and stored in a non sexual part of the body (i.e., big toe). The ego was impoverished, but retained the ability for a phantasized, albeit illusional, sexual activity. The psychotic episode may represent the patient's inability to embrace the physical change, the appearance of the syndrome and the operation as "demands on the psyche of the change of the body image" [17].

Conclusions

To the authors' knowledge this is the first case to provide evidence of a correlation between vaginal aplasia

(MRKH) and sexual delusion in a schizophrenic patient. Clinicians should be aware of this association and offer patients with MRKH psychiatric evaluation followed by psychotherapy if necessary.

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