The Phenomenology of Delusions in a Patient with Disorders of Sex Development

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ABSTRACT

Abnormal development of the external and internal genital organs and male pseudohermaphrodite-type disorders of sex development is one of the conditions that creates problem in determination of gender. In this case report, our aim is to discuss how disorders with psychotic symptoms may affect different cultural life styles, circumstances, experience, delusion contents of identification and acceptance in a patient diagnosed with

bipolar affective disorder, and with male-pseudohermaphroditism during adulthood.

Keywords: Bipolar affective disorder, disorders of sex development, delusion, phenomenology

INTRODUCTION

In order to maintain a healthy sexual development, installation of the sex chromosomes in a normal manner is required. It is known that many diseases are caused by numerical or structural irregularities of X and Y chromosomes. Abnormal development of the external and internal genital organs and conditions that cause a problem in sex determination are defined as disorders of sex development (DSD). One of these is deficiencies of androgen biosynthesis and metabolism leading to insufficient virilization in boys. 17 hydroxylase deficiency, 17 β -OH-dehydroxylase deficiency and 5 α -reductase deficiency are some of these (1,2). 17 hydroxylase deficiency is a rare disease where cortisol, androgen, and estrogen cannot be synthesized as a result of a defect in the synthesis of steroid. Such patients are usually raised as females, and when they grow older they are admitted to clinics with symptoms such as primary amenorrhea and hypertension (3). Additionally, defects in testicular development, resistance to androgens, and congenital adrenal hyperplasia may lead to distorted genitalia. Gender ambiguity of these children may be recognized after birth or later (2). Sexual identity development in individuals with ambiguous genitalia is an important dimension of the problem.

Delusions are unrealistic thoughts that cannot be resolved with a reasonable explanation and that do not fit into the recent era and society (4,5). It is not clear how the delusions, one of the most colorful tables that can be seen in mental disorders, occur; however, there are many opinions on this issue (6). In order to explain delusion, "conditioning and misperception" theories are proposed. According to the conditioning theory, patients are prone to create delusion due to their mental condition. These patients are trying to elude their troublesome conditions with the help of delusions. According to the misperception theory, delusions arise from the disclosure requirements of abnormal experiences, and sensory impairment in the patients' abilities distort their observations (6,7,8).

In this case report, our aim is to discuss how disorders with psychotic symptoms may affect different cultural life styles, circumstances, experience, delusion contents of identification and acceptance in a patient formerly diagnosed with DSD with male-pseudohermaphroditism and followed up with the diagnosis of bipolar affective disorder (BAD).

CASE

Miss D was a female, 35 years old, single, high school graduated patient with completely feminine outlook, and she was not working. She was willingly admitted to a polyclinic, accompanied by her brother. She reported that she was confused about her gender. According to her anamnesis, she was raised in a culture where boys and masculinity function are important, she has five brothers, her five sisters who were born before her died before the age of 3 due to unknown reasons. In 2002, she was admitted to the endocrinology unit of a university hospital with complaints of amenorrhea, elongation at height, growth in hands and feet. As a result of the examinations, she was diagnosed with "male pseudohermaphroditism (17-hydroxylase deficiency)." During this period, prednisolone 5 mg/day was administered due to adrenal insufficiency, and she has been followed-up in the clinic at regular intervals. The patient was raised according to the female gender by her family, and she also adopted this gender. She reported that when she learnt that her gender was not clear and her chromosome structure was 46 XY in 2002, she got confused and hesitated even when wearing clothes. She said she feels like a woman, she knows that she looks like a woman; however, when the doctors said "your gender is not clear, your chromosome structure is that of a male but you look like a woman," she got confused. Additionally she said, she doesn't know how such patients behave and according to which gender they dress.

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According to her psychiatric history, she suffered from depression in 1999, but recovered without any treatment, and she had manic attack in 2000 and was diagnosed with BAD. Although she used to wear a turban, she decided not to wear it and started applying make-up and talking excessively. After some time, she started to hear a voice saying "You are special; we will take you with us." She believed that she was more important than others, and she was the first woman chosen to save the world. During that period, she was hospitalized with the diagnosis of BAD manic episode in a university hospital for a month. She was discharged with remission and her medication therapy included valproic acid 1000 mg/g and quetiapine 400 mg/g. After a year, she had complaints of not enjoying life, unwillingness, sleeplessness, and she was diagnosed with depressive episode, so her medication therapy was rearranged as valproic acid 1000 mg/g, fluoxetine 20 mg/g, and quetiapine 400 mg/g. when she was diagnosed with male pseudohermaphroditism, she was followed-up in the same hospital with the diagnosis of depressive episode. During that period, depressive symptoms were thought to be triggered by the corticosteroid she was taking, so endocrinology consultation was requested. But the endocrinologist did not alter the dosage. Her psychiatrist in the same hospital rearranged fluoxetine dosage to 60 mg/g. Her depressive symptoms improved, and her treatment included valproic acid 1000 mg/g, fluoxetine 60 mg/g, quetiapine 200 mg/g, and prednisolone 5 mg/g. In 2005, her father passed away; she started to have complaints of sleeplessness, excessive talking, and restlessness. When she had grandiose delusions such as "I am an angel; therefore, I don't have a gender, because angels do not have a gender," "I am superior to other people" and psychomotor agitation, she was hospitalized for one and a half month. Fluoxetine was gradually discontinued and risperidone 4 mg/g was added to treatment. When she was discharged, she still had the delusion of "I am an angel because I don't have a gender," but it gradually disappeared with time.

During her mental examination, she was conscious, oriented, and cooperative, her mood was euthymic, her affect was anxious, and she was extremely engaged in thinking about which gender she had. There weren't any psychotic findings. She didn't have suicidal ideas. Her judgment was realistic, and she had insight. According to her family history, her big brother also had BAD. She was still taking valproic acid 1000 mg/g, quetiapine 400 mg/g, and prednisolone 5 mg/g. Psychotherapy was added to her treatment and polyclinic follow-up was planned at regular intervals.

DISCUSSION

It is not clear yet how the delusions, one of the basic characteristics of mental disorders with psychosis, are formed. According to some studies, high doses of dopamine are required to maintain delusions caused by increased-dopamine-affected brain functions. In fact, in a study in patients with delusions of being harmed and erotomania, it was found that patients' homovanillic acid levels and dopamine metabolites were higher than those in the normal population (9,10).

However, the content of delusions were reported to be affected by the patient's socio-cultural and socio-demographic characteristics (11,12). Life experiences of individuals can affect the nature of the psychological conflicts. Therefore, clinical images of psychiatric disorders are influenced by individuals' structural predispositions, personality traits, and perception of stress (13). Our patient was diagnosed with BAD when she was 23 yrs and with male pseudohermaphrodite-type DSD when she was 25 yrs. In hermaphrodite cases, it is reported that those who were raised in accordance with the female gender are diagnosed in older ages. Our patient being diagnosed at the age of 25 yrs supports the information obtained from literature (14). Our patient adopted the female gender and sexual identity until she was diagnosed with hermaphroditism. She had her first manic episode when she was 23 yrs, and had grandiose delusion of being the first woman chosen to save the world. It was reported that the content of her grandiose delusions during the manic episode were similar till she was diagnosed with hermaphrodite. In manic patients, grandiose delusions

such as physical strength, status, talent, wealth, and discovery-invention are common. It is suggested that these delusions are formed in response to stress for being protected against external events or distressing mental cognitions, and have a protective function. Therefore, their vulnerable and weak ego is protected from distress and forms a false unconscious self (15). As our patient is from a male dominated culture, it can be said that to get rid of feelings of worthlessness due to the female gender, our patient protected herself from distressing cognition with the grandiose delusions of being "the first woman chosen to save the world."

The content of the patient's grandiose delusions changed after being diagnosed with hermaphrodite. Grandiose delusions such as "I am different from others, I am an angel because angels do not have gender" reflecting confusion arising from the uncertainty of the gender support the idea that the content of delusions are affected by different condition, experience, and acceptance. Maher suggests that delusions arise as a result of normal reasoning made to explain abnormal perceptual experiences (16). In light of this suggestion, we may consider that our patient's distressing thoughts about the uncertainty of gender turned into abnormal perceptual experiences during the manic episode. It can be considered that she is trying to avoid this distressing mental cognition by concluding through normal reasoning that "she is an angel because angels do not have gender."

As studies on contents of delusion are few in literature, it is difficult to determine the individual, social, and cultural differences of the delirium content. We discussed how different cultural experience, conditions, experience, identification, and acceptance may affect the content of delusion. We believe that new studies performed with larger samples or larger study groups may lead to a better recognition of the disease by profiling delusions, and can contribute to the development of new treatment options.

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