Remission of Episodic Sweating Attacks and Comorbid Depression in Shapiro Syndrome: Case Report

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ABSTRACT

Shapiro syndrome, a rare disorder originally described by Shapiro and Plum in 1967, is characterized by episodic hypothermia and hyperhidrosis associated with agenesis of the corpus callosum. Proposed hypotheses to explain the clinical features of this syndrome include changes in the set point of the hypothalamic thermostat, increased norepinephrine (NE) release, and decreased plasma NE clearance. It was emphasized that the recognition of Shapiro syndrome in the evaluation of episodic hyperhidrosis is important. Here, we described a case with Shapiro syndrome who presented to our psychiatry clinic with recurrent episodic profuse sweating and depression. Sweating attacks and depression remitted after successful treatment with amitriptyline.

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Key words: Shapiro syndrome, corpus callosum agenesis, hyperhidrosis

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Introduction

Shapiro syndrome originally described by Shapiro and Plum in 1967, is an infrequent disorder. It is characterized by episodic hypothermia and hyperhidrosis associated with corpus callosum agenesis. The hypothesis proposed to explain the change of hypothalamic thermostat sets the result of increased norepinephrine (NE) release and decreased plasma NE clearance. In previous studies, it was emphasized that the recognition of Shapiro syndrome in the evaluation of episodic hyperhidrosis is important. Time period between two attacks may be either hours or years, and attacks may take either hours or years to occur. Until now, only 30 cases are reported in the English literature. Certain pathophysiological mechanisms were not clear for the syndrome. The accepted mechanisms include the set point of decreased hypothalamic thermostat, NE and melatonin dysfunction, and possible inflammatory periods that affect epileptic focus. Here, we describe a case of a patient with Shapiro syndrome visited to our psychiatry clinic with recurrent episodic

profuse sweating and depression episodes. Sweating attacks and depression remitted after a successful treatment with amitriptyline.

Case

Complaints: A 31-year-old, primary school graduate housewife who was married and had one child visited our psychiatry hospital with sweating, tremor, fatigue, malaise, and reluctance attacks.

History of disease: She had sweating and tremor attacks for 14–15 years. She initially visited an external neurology clinic in 2007; cranial magnetic resonance imaging (MRI) is planned, and corpus callosum (CC) agenesis is detected. However, the patient was referred to our psychiatry department without any diagnosis. In 2009, she started to feel infelicity, reluctance, malaise, anhedonia, lack of appetite, and sleeplessness. The at-

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Figure 1. Sagittal MRG

tacks lasted for half an hour and occurred 3–4 times a week. Finally, she visited Erenköy Psychiatry Hospital firstly in 2009 and presented with fatigue, lack of appetite, anhedonia; secondly, in March 2010, she presented with sleeplessness, reluctance, and refusal to eat and drink following a miscarriage. She had been treated for the diagnosis of mixed anxiety and depressive disorder, and she was discharged up to two times with the consent of her family members. In June 2010, she was admitted to the psychiatry clinic again with severe sweating as well as tremor attacks and unhappiness. During the clinical observation with mixed anxiety and depressive disorder diagnosis, she had also been diagnosed with Shapiro syndrome with CC agenesis associated with hyperhidrosis, as indicated by the test results. During hospitalization, the patient was treated with mirtazapine for sedation initially with 15 mg and then 30 mg/day of alprazolam for anxiety and 1 mg/day of sodium valproate for possible epileptic seizures with recommendation of clinical neurologist as 1000 mg/day. She was discharged after decreasing depressive symptoms with partial remission of hyperhidrosis. Some psychological tests during hospitalization, namely Rorschach, Alexander, and Bender–Gestalt tests were administered. There was no pathology on EEG report. Alexander test IQ was 80, indicating borderline mental capacity and Bender–Gestalt test was “significant in terms of organization.” The result of Rorschach reported poor ideal content, stereotypical thinking style, away from interpersonal relationships, and weakness of social cohesion. Patient without regular outpatient follow-up and drug use finally stopped the medication completely in October 2010 because of pregnancy planning again. In September 2011, former drug therapy was started again by readmission to our hospital on recurrence of the same symptoms. For sweating attacks and

Figure 2. Axial MRG

electrolyte imbalance, fluid replacement therapy was started and the patient was discharged with partial remission. Although, family was educated regarding treatment and disorder, the patient could not use her drugs after discharge.

Personal history: No other disorder, no history of smoking, no drug use, no suicide attempts, and unexpected abortion of 4-month-old fetus in 2011.

Family history: There was nothing noteworthy.

Mental status examination: The patient had moderate self-care and decreased psychomotor activity in collaborative attitude. She was depressed and anxious, and she had decreased speech rate and spontaneity. The flow of thought was normal, and associations were regular and purposive. She felt that she was worthless and incompetent; however, she did not have thoughts of suicide or homicide. She had rough cognitive functions, and she had sufficient internal reasoning and insight. Physical examination findings were assessed as roughly normal, except excessive sweating and very light hypothermia (36°C). Patient’s MR imaging with axial, sagittal, and coronal sections are shown in Figures 1,2,3.

Clinical observation: In 2012 May, the patient was admitted to our psychiatry clinic again with recurrent unhappiness, malaise, sleeplessness, excessive sweating, and tremor. For her depressive symptoms and hyperhidrosis, amitriptyline was started at 50 mg/day and increased up to 100 mg/day. After neurological consultation, valproate of 1000 mg/day was planned to be continued. Tremor attacks and depressive symptoms disappeared

Figure 3. Coronol MRG

in 1 week. Hamilton Depression Scale score decreased from 14 to 4. There was no pathological finding in weekly biochemistry tests. The patient was discharged with available drug therapy and psycho-educational suggestions. For a period of 4 months, the patient's symptoms did not reappear, and medication compliance is better in the outpatient follow-up.

Discussion

The CC is the largest nerve fiber that connects the two hemispheres. The basic function of CC is providing hemispheric integration in relevant areas by establishing a link between the two hemispheres. CC generally serves to transfer sensory information similar to a conduit. In CC agenesis cases, most of the symptoms that affect the patients who underwent an operation for the separation of the two hemispheres are not observed. That result is associated with alternative pathway like that provides the link between hemispheres and anterior commissure that remains mostly healthy in CC cases. Other possible compensatory mechanism may be that some specialized functions, such as speech, are duplicated in both the hemispheres.

The most common clinical presentation of Shapiro syndrome is hyperhydrosis and hypothermia attacks related to CC agenesis. In prior studies, it is emphasized that sweating attacks asso-

ciated with hypothalamic dysfunction are important in Shapiro syndrome. A study by Darku B. et al. in 2011 indicates that clonidine therapy is effective to control these attacks. In our case, to control depressive symptoms or symptomatic treatment of sweating attacks, amitriptyline is gradually increased up to 100 mg/day. During the 4-month clinical follow-up, symptoms and complaints are not recurrent, indicating that the patients with Shapiro syndrome can be treated in this manner.

The comorbidity of some clinical patients, such as learning difficulties and epilepsy, is commonly observed with CC agenesis. The patients with learning difficulties, low self-esteem, and lack of self-confidence stand out as elementary causes in treatment failure. In our case, the patient was an average student in primary school and could not continue education; in addition, she has borderline mental capacity, showing lack of self-confidence, as included in the literature.

In the literature, some clinical patients having anxiety symptoms can lead to psychosis. Approximately 40% of the patients with CC agenesis have epilepsy that began in childhood or adulthood. High frequency of seizures in agenesis reminds the inhibitory nature of CC structure. Although most of the published cases were analyzed to examine an obvious neurological syndrome, the significant rate of some case series (approximately 13%) is asymptomatic in terms of neurology. In our case, any positive symptom suggestive of epilepsy is not observed in general neurological assessments and on EEG records.

Our report showed that Shapiro syndrome should be taken into account in patients with excessive sweating attacks and amitriptyline may help these patients in the control of their symptoms.

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