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CASE REPORT

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Loss of Sight Caused by Ocular Self-Mutilative Behaviour: A Case of Malignant Tourette Syndrome

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ABSTRACT

Tourette's Syndrome is developmental neuropsychiatric disorder characterized by stereotypic, non-rhythmic multiple motor and/or vocal tics. In rare cases, severe tics which can be life-threatening or self-mutilating may be observed in Tourette Syndrome. These types of cases that involve severe self-injurious behavior are called malignant Tourette's Syndrome. In this report, we present an adult case of Tourette Syndrome with vision loss as a result of recurrent and severe ocular self-mutilative tics.

Keywords: Tourette's syndrome, self-mutilation, vision loss, adult

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INTRODUCTION

Tourette Syndrome (TS) is described as a developmental neuropsychiatric disorder generally characterized with stereotypical, nonrhythmic, multiple motor or vocal tics (blinking, sniffling, clearing one's throat, neck and shoulder movements etc.) (1). Starting in the childhood, while the disease is mostly observed in children below age 18, disease symptoms can continue into the adulthood (2). In rare cases, severe tics, which are presented as life-threatening and self-mutilative level of injuries, may accompany TS. Cases including these types of severe self-mutilative behavior are named as malignant TS, and the rate of malignant TS has been reported as 5% among all TS cases (3). Self-mutilative behaviour such as hitting the head, punching/slapping the body, punching/slapping the face or hitting their body with hard objects are observed often in malignant TS cases, and ocular self-mutilative behaviour is observed very rarely (4). In this letter, an adult TS case with repetitive ocular self-mutilative behaviour is presented.

CASE

The male patient, who was 53 years old, primary school graduate, married and lived with his family has been accepted to emergency department of Erenköy Mental Health Training and Research Hospital, where he was directed for consultation from the hospital he was receiving inpatient treatment, namely Göztepe Training and Research Hospital Eye Diseases Service. From the consultation note, it was learned that the patient damaged his left eye by involuntary actions of applying pressure with his finger, and that he has been operated 15 days ago due to the development of retinal detachment caused by giant retinal tear. In addition, the patient continued involuntary damaging behaviour with his finger on his left eye after the surgical intervention, and he was operated a second time due to developing retinal detachment in his left eye. Since patient's involuntary harming behavior continued on his left eye after the second operation, a protective *'eye-shield'* was placed on the left eye, and the patient was directed to us for regulating treatment.

According to the information received from the patient and his family, he had complaints of head jerking, involuntary movements such as damaging his eye by pressuring with his finger, yelling "Don't!" after these movements, making incoherent sounds, and difficulty in falling asleep. It was learned that the tics first started in adolescence with coprolalia and vocal tics, afterwards complex motor tics were included such as breaking and throwing objects (watch, remote control, glasses etc.). It was also learned that ocular self-mutilative behaviour first started 3 years ago by applying pressure on his right eye with his finger, and that he developed total vision loss in his right 1 year ago since these uncontrolled movements caused retinal detachment. After developing vision loss in his right eye, the patient started damaging his left eye similarly with involuntary finger movements. It was learned that the patient had applied to psychiatry various times after his complaints that appeared in adolescence, however he did not receive regular treatment, and he never showed complete improvement while the severity of his vocal and motor tics varied from time to time. He had applied to psychiatry 3 years ago due to ocular self-mutilative behaviour, and he used various selective serotonin reuptake inhibitors irregularly.

In the first psychiatric examination of the patient in emergency department, he was conscious, cooperative and had full orientation. The patient could perform self-care, while his mood and affection was depressed. He had normal speech rate, and associations were regular and goal-oriented. No hallucinations or delusions were detected. He had no obsession, phobia and mental overexertion in his thoughts. No suicidal or homicidal ideation was determined. He had insight. His memory function was protected, knowledge and intelligence level was sufficient,

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and the patient was observed to have motor and vocal tics consisting of head jerking, applying pressure on the 'eye-shield' placed on his left eye in order to damage the eye, and making incoherent sounds during the interview. No neurological pathology was determined in the neurological consultation of the patient, and cranial MRI imaging was evaluated to be within normal limits. It was learned that he received an additional diagnosis of major depression 3 years ago when he applied to psychiatry for his self-mutilative behaviour on his right eye with involuntary finger movements. It was learned that 20 mg/day fluoxetine tablet was started lastly for major depression treatment and he continued to use them. Since the speech, knowledge level, vocabulary, alignment of events, abstraction and judgment of the patient were evaluated during the interview and it was concluded that his intelligence level was sufficient, no mental deficiency or limited mental capacity was determined. Obsessive compulsive disorder diagnosis was also excluded since he had no obsession and compulsions.

The drug treatment that the patient was using during his application consisted of 20 mg/day fluoxetine tablet and 2 mg/day risperidone tablet. The patient was followed-up once a week for four months with Tourette Syndrome diagnosis, and risperidone was gradually decreased and stopped since it was though he received no benefit from risperidone. In its place, 5 mg/day aripiprazole tablet was started and the dose was slowly increased up to 10 mg/day. The patient's depressive symptoms were insignificant during application, but fluoxetine tablet dose increased up to 40 mg/day since increased depressive complaints were observed in the second week following his discharge from the hospital. Depressive symptoms decreased by increasing the fluoxetine dose, and his tics continued albeit being reduced by approximately 50%. After that, 100 mg/day quetiapine tablet was started and its dose was increased up to 300 mg/day. At the end of the fourth month following his first application, patient's treatment consisted of 40 mg/day fluoxetine tablet, 10 mg/day aripiprazole tablet and 300 mg/day quetiapine tablet. Although tics such as head jerking and making incoherent sounds continued mildly in control examinations, generally the patient was observed to have significant decrease in vocal and motor tics. Throughout the control period, it was learned that he did not continue applying involuntary pressure on his right eye with his finger. In addition to total vision loss in his right eye that was present before the application, it was noted in the control evaluation performed by the ophthalmologist that severe vision loss (95% vision loss) continued in his left eye.

DISCUSSION

In recent articles, tics in Tourette Syndrome are observed to be classified under tonic, clonic (jerk-like), dystonic (sustained), and blocking (stopping movement or speaking) tics (5). In addition, tics are classified as 'simple tic' and complex 'tic' according to involving one or multiple muscle groups (6). Simple motor tics only involve a muscle group, and causes short, jerk-like movements. While simple tics often start and end abruptly (clonic tic), it may sometimes cause a short abnormal posture (dystonic tic) or isometric contraction (tonic tic) (6). Blinking, head jerking or palatal myoclonus may be examples of simple clonic tics (7). Simple dystonic tics include movements such as blepharospasm, eye movements, bruxism, keeping the mouth open, torticollis and shoulder rotation. Simple tonic tics can be presented as contracting stomach and leg muscles without moving (8). Complex motor tics are presented as consecutive and coordinated uncontrolled movements of more than one muscle group generally resembling normal activities, or uncontrolled complete words and sentences. Repeated touching, throwing, hitting, jerking, jumping and body bending movements are the most common ones of complex motor tics. Copropraxia, which is presented as obscene gesturing or touching another person's genitalia, repetitive vomiting, belching or air swallowing are rarely observed motor tic examples. Complex motor tics sometimes can be observed as movements that harm the individual's own body (9). Apart from other complex tics such as breaking and throwing objects, our case had involuntary clonic finger movements that harmed his own eye. Complex motor tics can sometimes be confused with compulsions in obsessive compulsive disorder in clinical practices. The fact that our case did not have obsessions in his thoughts, had involuntary motor movements, his behaviour was not organized and not goal-oriented shows that ocular self-mutilative movements are complex motor tics that is a part of TS.

Being descriptive and characteristic for tic disorders, TS is a neuropsychiatric disease that starts in childhood and involves several motor tics and one or more vocal tics together. Although it was indicated in medical history of our case that the tics appeared in adolescence, it is highly possible that especially simple tics could have been overlooked, and it was considered that tics had started in childhood and became more severe in adolescence. It has been reported that tics are self-harming in 33%-60% of TS patients (4, 10). TS cases with self-harming tics are named as malignant TS (3). In this respect, our case can be considered to have malignant TS due to his severe self-harming behaviour towards his own eye. It has been suggested that motor and behavioral symptoms in TS is associated with the disruption of impulse control, and the normal inhibitor mechanisms on primitive behaviour is decreased (11). While its pathogenesis is not completely understood, it has been indicated that movement disorder in TS is caused by the disruption in cortico-striatal-thalamo-cortical pathways that play a role in motor control; and tics presented in TS are caused by the insufficiency of the cortical inhibition on the involuntary motor movements induced by the activity of basal ganglia (12). While the reason of the disruption in cortico-basal ganglia activity is not completely understood, it has been suggested that anomalies in γ-aminobutiric acid (GABA) system contributed in the disinhibition in TS (5). In addition, the abnormal plasticity in motor cortex, basal ganglia and brain stem has been suggested to play a role in motor control deterioration in TS (5). It has been reported in a neuroimaging study that gray matter thickness is decreased in precentral, postcentral and superior, inferior and internal frontal sulcus areas and this decrease is associated with the severity of the tics (13).

In a recent meta-analysis study, it has been reported that the efficacy of aripiprazole in TS treatment is similar with haloperidol, tiapride and pimozide, and it is more advantageous compared to these drugs with regard to extrapyramidal side effects. The treatment dose of aripiprazole has been suggested as 10 mg/day in this meta-analysis study (14). After seeing in our case that risperidone has very limited effect on tics, it was switched to aripiprazole, and after obtaining sufficient response with 10 mg/day aripiprazole, the treatment was switched to combination treatment, which is the next step according to the recommendations of European clinical guidelines for Tourette syndrome and other tic disorders (15). Upon considering the complaint of insomnia which appeared from time to time, 100 mg/day quetiapine was started and it was gradually increased up to 300 mg/day. Furthermore, fluoxetine dose was increased to 40 mg/day due to his depressive complaints that appeared two weeks after discharge.

Similar to our case, there is only one case reported in literature that applied pressure on the eye with their finger as a component of tics in TS and thus caused retinal detachment (16). Our case has lost 100% of his sight in his right eye and 95% of his sight in his left eye due to retinal detachment, and has experienced a near complete vision loss. It was observed in literature that severe ocular self-mutilative behaviour is mostly explained by schizophrenia, dementia or mental retardation (17, 18). Apart from those, ocular self-mutilative behaviour observed in psychiatric disorders such as obsessive compulsive personality disorder or borderline personality disorder is not severe enough to cause

blindness (17). In this case series, it should be considered that TS may be overlooked. Our case shows that self-mutilative behaviour observed as a part of tics in malignant TS may be severe enough to cause vision loss. While our case only had depression as additional diagnosis, the possibility of comorbidities, especially high possibility of accompanying intellectual developmental disorder and OCD spectrum disorders should be considered in malignant cases. Apart from these disorders that are mentioned during the psychiatric consultation of cases determined to have ocular self-mutilative behaviour, it is also important to consider TS, which rarely continues in adulthood. Furthermore, upon considering the treatment of our case that also had an additional diagnosis of depression, it is concluded that obtaining positive results with only SSRIs and antidopaminergic treatment without using medicines such as clonidine and guanfacine is an important result particularly for TS accompanied by depression.

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