Tourette's syndrome with rapid deterioration by self-mutilation of the upper lip

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A case of Tourette's syndrome is presented in a thirteen-year-old boy with obsessive-compulsive symptoms including self-mutilation of the upper lip. His upper lip injury was caused by complication of picking with fingernail, and self-biting with the lower anterior teeth. It became rapidly worse and the median part of the upper lip collapsed. But the placement of an acrylic splint was able to prevent further damage of the upper lip.
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INTRODUCTION

A tic is defined as involuntary, sudden, rapid, recurrent non-rhythmic, stereotyped motor movement or vocalization and is divided into four groups: transient tic disorder; chronic motor or vocal tic disorder; combined vocal and multiple tic disorder (Tourette's syndrome); and tic disorder not otherwise specified.[1] Gilles de la Tourette's syndrome (Tourette's syndrome, TS) is a chronic, familial neuropsychiatric disorder that is characterized by the presence of multiple motor and one or more vocal tics at some time during the illness.[1,2]

It develops during childhood between the ages of two and thirteen, affecting males more than females, and often begins with a involuntary twitching (simple or complex motor tics) of the muscles in the face.[2] Subsequently TS shears vocal tics, which include explosive involuntary utterances, coughs, grunts and echolalia. When the patient is over ten years old, coprolalia with or without echopraxia and copropraxia may exist.[2,3]

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The tics occur many times a day recurrently throughout a period of more than one year. During that period, there is never a tic-free period of more than three consecutive months.[1] Motor tics of the oral region include facial grimacing, licking of the mouth and cheek, lip smacking, jaw snapping, coughing, bruxism, biting the lip and cheek, and picking of the oral tissue using the fingernail.[4-7]

It has been well known that obsessive-compulsive symptoms are present 10% to 90% of the patients with TS.[2,8-10] Obsessions include intense intrusive, unwanted thoughts, such as concerns about bodily wastes and secretions, unfounded fears, need for exactness, symmetry, evenness, and neatness, excessiveness, perverse sexual thoughts, and intrusions of words and phrases.[3,10,11] Compulsions are composed of the subjective urge to repeat meaningless and irrational rituals, such as checking, counting, cleaning, washing, touching, smelling, hoarding, and rearranging.[3,10,11] About 5 to 50% of patients with TS have an aggressive behavior such as self-mutilation, which is supposed to be a compulsive tension-releasing mechanism with a self-injury of oral tissue often observed.[2,6,8,11-16]

The drug therapy is a treatment of choice for TS and haloperidol and pimozide which are the neuroleptic agents (dopamine D_2 receptor antagonist) that have been used.[17] In some cases, when the anxiety is severe, it is effective to use anxiolytics as well.[2]

When the obsessive-compulsive symptoms are found, clomipramine (tricyclic antidepressant) or fluoxetine (selective serotonin reuptake inhibitor) are prescribed.[18] Needless to say, psychotherapy can be effective and some slight cases have been improved by it.[1]

There are rare cases that may become dementia or schizophrenia for the prognosis of TS. On the contrary, there are many cases in which the symptoms will improve or even disappeared spontaneously during

adolescence and adulthood, and often the patients are able to live good social lives even though they have some tics all their lives.[2] The problem is that complications like obsessive-compulsive symptoms and impulsive behavior make the social and occupational functioning difficult.[1,2]

In this article we present a case of Tourette's syndrome (TS) with self-mutilation of the upper lip, which rapidly worsened and resulted in a defect of the median portion of the upper lip. We briefly discuss the oral self-mutilation of TS.

CASE EPORT

A thirteen-year-old boy was consulted in the oral surgery clinic to evaluate the non-curing stomatitis of the upper lip. He first showed muscle twitching of his body when he was eleven years old. After that he arose with curious actions such as covering his face with a towel, inserting a handkerchief into his nostril, touching his mouth with his fingers and licking his mouth all over with his tongue. His parents thought these actions were a habit and that there was no serious problem.

But, when he was twelve, he had complained of severe oral pain, and then he was admitted the local general hospital for severe stomatitis of the upper lip. After that time, though he had been admitted to the hospital twice, he had not consulted the psychiatric doctor.

Two months before it had been prominent to give a cough and to clear the throat, and he could not give up them though his mother cautioned him. Five days before he began to show a fine uncontrollable twitching of the body with self-biting of his upper lip and he could not go to school anymore.

Three days before, when his mother got very angry with him about his behavior, his condition got worse and he began to cry and complain of the severe oral pain.

The patient lived with his parents and two siblings and they were all in good health. There was no familial occurrence of psychiatric disorder including TS. He had been sucking his finger from his childhood and had enuresis till he was eleven years old. He was not a problem child and he got average grades at school.

On physical examination he was well nourished and healthy, but a muscle twitching on his face was found during his consulting. On local examination, there was a relatively large ulcerative stomatitis with pain around the labiogingival junction of the upper lip (Figure 1) and also it was discovered that his fingers picked the inside of the upper lip repeatedly. As the psychiatric cause was strongly suspected for his disorder, he was consulted the by psychiatric doctor.

On psychiatric examination, he showed involuntary movements (tics) of his face, especially the stretching of his upper lip vertically, with a sudden twitch of the trunk. Vocal tics and inarticular noises (throat clearing) were found, but coprolaria was not found. During the examination he repeatedly wiped around his neck and face with his hand or a towel compulsively.

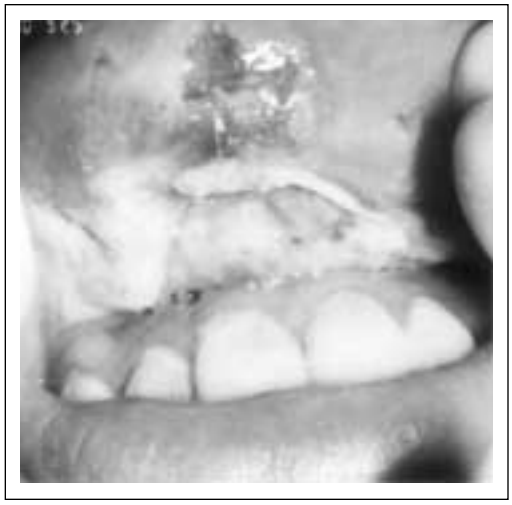

Figure 1. Ulcer caused by picking with his fingernail is found along with labiogingival junction of the upper lip.

He was diagnosed with a suspicion of Tourette's syndrome by the psychiatric doctor. As the psychiatric clinic did not have a ward in our hospital, he was admitted to our clinic to relieve the stomatitis and to continue the psychiatric examination.

He was given antibiotics and haloperidol 3mg t.i.d., and bromazepam 6mg t.i.d. and fulnitrazepam 1mg h.s. were also given from the next day.

He showed motor tics and vocal tics repetitively. The motor tic was prominent in the muscle in his face, neck and shoulder. Vocal tics included explosive involuntary utterance, however articulate obscenities were not found.

On the second day, though he continued to touch his mouth with his hands and was often chewing tissue paper, however the stomatitis appeared to be improving. EEG and MRI, resulting from the consultation with the pediatric doctor, were within normal limit.

On the sixth day, the tics and the obsessive-compulsive syndrome were getting worse, while hitting his head by himself with crying newly arose, and verbal echolalia was prominent. As he began to bite himself in the upper lip tightly with his lower anterior teeth and as the picking with his fingernail at it was persistent, the stomatitis got rapidly worse again. He complained greatly that if he did not touch his upper lip though it was painful to do so, he was seriously anxious. We warned him that the stomatitis would get worse by his picking and biting of the upper lip. However trying to control the matter was not easy and he continued self-mutilation in agony.

Haloperidol was increased to 6mg, but was relatively ineffective, so it was reduced to 3mg. An injection of diazepam at 5mg, which was to some degree effective even though in short period, was added intravenously to sedate him.

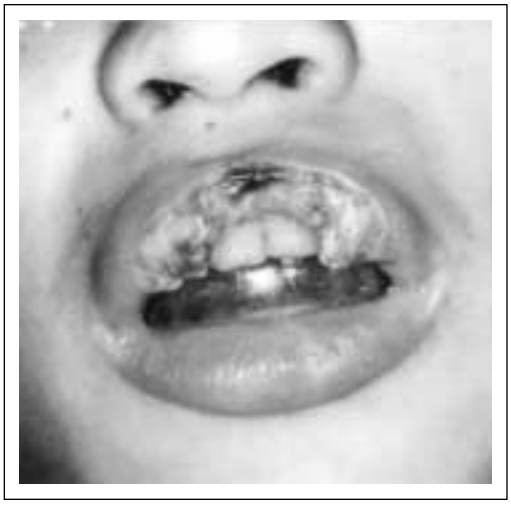

Figure 2. Median portion of the upper lip is defected and the surface is covered with necrotic tissue. The acrylic splint is placed in the mandible.

On the ninth day, as the obsessive-compulsive syndrome became thoroughly worse, he also broke his legs on the bed frame. Gradually the ulcer became deeper and more destructive, and deterioration of the upper lip became evident. The psychiatric doctor was permitted to temporarily restrain the patient in the daytime, but only the restraint of his legs was intermittently continued on the eleventh and twelfth days.

On the thirteenth day, the number of tics decreased and the obsessive-compulsive symptoms slowed down, the impression of the mandible was made after obtaining his consent.

On the fifteenth day, he agreed to accept an acrylic splint on his mandibular teeth because he realized that the splint alleviated his pain (Figure 2). After this, the tics and the compulsion to touch his mouth were still evident, however self-biting of the upper lip by the mandibular anterior teeth had disappeared and the destruction of the upper lip stopped. Since then he voluntarily continued to use the splint because he seemed to be afraid of the pain that would reoccur by removal of the splint.

On the twenty-first day, after the diagnosis of Tourette's syndrome, he was discharged from our clinic for further treatment in another psychiatric clinic.

DISCUSSION

As a patient with a self-mutilation may initially visit the dental or oral surgery clinic, we should keep in mind that self-mutilation is caused not only simply by frustration, boredom and tension, but also by schizophrenia, personality disorder, Lesch-Nyhan syndrome, Cornelia de Lange syndrome, fragile X syndrome, neuroacanthcytosis, obsessive-compulsive disorder, TS, and other neurobehavioral disorders.[2,3,11] It is important that the diagnosis is made by the psychiatric doctor after careful examination when the cause of the disorder is questionable. In this case, it was not until the consultation with the psychiatric doctor was finished after visiting our clinic for severe stomatitis that the patient was diagnosed with TS.

With regard to self-mutilation, though the patients are aware of the pain that follows self-mutilation, they cannot stop to get a brief relief from the compulsion.[2,7] The self-mutilation of the oral tissue in TS included self-biting of the tongue, lower lip and buccal mucosa, tooth extraction, and soft tissue ulceration using a fingernail.[2,6,8,11-16] It was rare to find biting of upper lip like in our case. We assumed that it was not easy to bite the upper lip by oneself.[19] Our case had the complication of picking with fingernail, which seemed to trigger the self-biting of the upper lip.

Generally, in the treatment of oral self-biting except for behavioral and pharmacological therapy, the oral local therapy includes padded blades, mouth props, an acrylic or metal splint, a tongue stent, intermaxillary fixation and the extraction of the teeth.[6,20-24] In cases of TS, as the improvement of self-biting is able to be expected along with the remission of the compulsion, the conservative treatment is desirable. At the same time, as no intellectual deterioration accompanies the disease in general,[2] it is important to obtain the consent of the patient before treatment, although it may take time.[6] In cases where the consent was obtained including our case, the patients accepted the oral therapy and the healing was good.[6,20]

But Lowe[6] described a case in which the splint was placed on the patient before his consent and the patient refused to accept the splint and removed it even though it had been attached by wires. Intensification of sedation may be effective, but it is only temporary treatment,[6] when touching or hitting by hand, or picking with a fingernail is the cause of the self-injury of the oral region, restraint may be an effective therapy to prevent an injury. However, it should be performed with the agreement by the psychiatric doctor because as the restraint might be getting worse, the psychiatric condition and the self-injury of another portion might occur even though the initial self-injury improves.[2] In our case only the restraint of the legs was performed to prevent the leg injury.

It is difficult to prognosticate the occurrence of oral self-mutilation in TS, but whenever the stigma of oral tics arises, prompt dental care should be planned as soon as possible. Sometimes self-mutilation of the oral region might suddenly arise and spread, and when so, it is impossible to obtain the consent to perform a dental approach such as taking an impression till the symptom improves. Our case, unfortunately, required reconstruction, through esthetic surgery for the sudden exacerbation of the self-biting of the upper lip.

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