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A case of Tourette syndrome presenting with oral self-injurious behaviour

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Summary. Self-injurious behaviour (SIB) is deliberate harm to the body without suicidal intent, and the condition occurs in a number of psychiatric, behavioural and developmental disorders. This case report describes a 4-year-old female with SIB who presented to a paediatric dentist after the self-extraction of teeth as a result of oral motor tics. The girl repetitively ground her teeth in a monophasic lateral motion that resulted in luxation of her maxillary right primary canine, and produced generalized oral and facial pain. The parents consulted the dentist about their child's complaint of toothache. The oral findings were unexcephonable except for a mobile primary canine, but there was a history of unusual behaviour including hyperactivity, and after multidisciplinary consultation and exclusion of other systemic diseases, the subject was diagnosed as suffering from Tourette syndrome (TS). Preventive treatment using a dental splint was provided. Noncontingent reinforcement therapy was successfully used to diminish the subject's SIB.

Introduction

Self-injurious behaviour (SIB) is deliberate harm to the body without suicidal intent [1]. The condition occurs in a number of psychiatric, behavioural and developmental disorders [2,3]. Oral SIB leading to tooth extraction can be seen in children who may scrape or pick at the gingiva with a fingernail. Compulsive touching and manipulation of the oral tissues may be a problem. If extreme, this may lead to extraction of permanent teeth [1]. The prognosis for the affected tooth or teeth depends on the amount of periodontal tissue that is destroyed. A form of self-mutilation to the teeth associated with nail-biting is the practice of sticking fingernail fragments intentionally into the gingival sulcus. Such a patient will present with gingival swelling of uncertain aetiology. This behaviour may be related to stress or anxiety, or it may be initiated in response to gingival irritation. If the child does not stop the habit or if multiple sites are seen, the dentist should consider referring the patient for psychiatric intervention [1].

Tics are the most frequent abnormal movement in children. These most commonly involve the orbicular muscle, and manifest as uni- or bilateral blinking, but tics may occur in any muscle group. It is common for tics to be transient and most authors consider transient tics to occur in many children during normal development. Tic disorders cover a wide spectrum, with Tourette syndrome (TS), also known as Gilles de la Tourette disease, being the most complex [4].

Tourette syndrome is a poorly understood condition that is characterized by repetitive tics, and utterances, noises or other phonic phenomena [5]. In 66% of cases, facial tics are the first symptoms. Reports indicate that SIBs occur in one-quarter to half of cases with TS [5–7]. Self-injurious behaviour in TS is correlated to the severity of the tic: milder SIB is related to obsessive/compulsive symptoms, while the more severe form is related to impulse/affect dysregulation, as has been postulated for SIB in other

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neuropsychiatric disorders [8,9]. Pain is a prominent and multifaceted feature of tic disorders [10].

Tics can cause social handicaps, especially coprolalia (involuntary swearing), but they may also be physically harmful. It is important for the dentist to recognize the early signs affecting the oral tissues in order to provide suitable preventive measures. Patients with tics consisting of jaw thrusting, repeated mouth opening, tooth-clicking or extreme nail-biting often develop orofacial pain and dysfunction of the temporomandibular joint [6,11,12].

Acts of orofacial mutilation include gouging of the eyes, pushing sharp objects into the ear canal, and lip-, cheek- and tongue-biting to the point of laceration [6,7,10,12].

This case report describes a subject who presented to a paediatric dentist with orofacial pain and SIB consisting of self-extraction of teeth resulting from oral motor tics.

Case report

A 4·1-year-old girl was referred to a specialist in paediatric dentistry by her general dental surgeon, who she had visited three times in previous weeks for toothache. The reason for referral was suspected toothache but without clinical evidence of caries or dental trauma. The parents described their child as having been screaming night and day for more than 2 weeks because of pain.

The medical history revealed that the child was under treatment with cephalosporin for impetigo that had promptly resolved. A specialist in otolaryngology found no other signs of irritation or cause for the pain. The parents reported no symptoms from the gastrointestinal system or problems with food intake. Pregnancy, delivery and psychomotor development had all been normal, and the child was considered to be generally healthy.

The subject is the younger of two sisters. The parents had noted that their daughter was hyperactive compared to her elder sister.

When questioned, the child said she had to bite. This gave her pain, but she reported that she still had to bite. Thus, the subject essentially described an obsessive–compulsive behaviour.

Oral findings

As a consequence of her hyperactivity, the child was not easy to examine. She did not adopt an attitude of reserve, as 4-year-old children often do at their first meeting with a new person. For example, when the subject was asked to show her teeth, she retorted that she wanted to look at the dentist's teeth instead.

The girl had a complete primary dentition with normal occlusion and with no obvious signs of caries. There was a slight hypomineralization of the teeth, but no significant attrition, suggesting that the grinding habit had started recently. The oral mucosa had a healthy colour except for the gingiva in the region of maxillary right canine, which was noted to be swollen, reddish and tender to palpation. The gingival sulcus around all the teeth was of normal depth. Several other teeth as well as the maxillary left canine were sensitive to percussion. The temporomandibular joints had a normal range of movement, but these as well as the masticator muscles were found to be tender to palpation.

From these findings, a disorder of mineralization resulting in abnormally sensitive teeth or an inflammatory disease of the joints and/or the gastrointestinal system were first suspected. A source of inflammation or tumours of the maxilla and/or mandible also had to be excluded. For diagnostic reasons, a panoramic radiograph was thought justified and was taken. This revealed no sign of dental disease or periapical destruction. The condylar anatomy also appeared normal.

Analgesics with an anti-inflammatory effect were prescribed as symptomatic treatment and had a positive initial response. The child stopped complaining of pain, but after a few days, she started screaming again.

The parents had noted that, since the toothache had started, their child was jumping instead of walking. She did not want to go outdoors and play with other children as before. It was obvious that the child was upset and did not feel well. The girl's limbs had no swollen joints, but the fact that she did not walk normally was thought to be a possible sign of a more generalized joint disease.

Therefore, the subject was referred to a paediatric rheumatologist who found no abnormalities except for the hyperactivity and the dislocation of a tooth.

At the next dental examination, the maxillary right primary canine was extremely mobile and dislocated at 45° in a buccal direction. Therefore, the tooth was extracted and sent for histopathological examination. The analyses showed no signs of any abnormal mineralization, caries, attrition or fracture that could cause her pain. The contralateral tooth, the maxillary left canine, was also becoming mobile, and

at this stage, the mandibular primary canines, first molars and incisors had tilted in a lingual direction. An individually designed occlusal splint was manufactured to stabilize the occlusion and to relieve the pressure to the teeth.

Since there were no local findings which could explain the girl's behaviour and rheumatology had been negative, she was referred to a neuropaediatric team.

Neuropaediatric evaluation

An expanded neuropaediatric anamnesis disclosed that the mother had suffered from tics at a younger age, but was no longer receiving any treatment and was well. The father had had concentration and attention problems when he was of school age, but had not been diagnosed as suffering from any specific condition.

Based on the history, physical examination, and follow-up, a preliminary diagnosis of Tourette syndrome was established.

Further questioning at this stage elicited that the subject had suffered from various tics for many months. These tics would disappear during sleep and could sometimes be suppressed for a time.

There were no other pathological findings, but on subsequent follow-up examinations, she had added the symptom of swearing that persisted for several months.

Over the period of review, the child has shown a variety of compulsive-obsessive behaviours and actions which have affected aspects of her daily life, including her education and social functioning. She now receives specialist help to try to overcome her difficulties.

Discussion

Tourette syndrome is a disease that is characterized by chronic motor and phonic tics. It is often accompanied by a variety of behavioural problems such as obsessive–compulsive disorder and attention deficit hyperactivity disorder, which may precede the onset of tics [4,5].

Tics are involuntary, sudden, repetitive, stereotyped movements or vocalizations which may wax and wane, and tend to involve the face. They may be exacerbated by anxiety and may be briefly suppressed voluntarily. Isolated tics and those included in behavioural disorders should be differentiated from stereotypes and disorders causing tics as secondary phenomena, such as encephalitis, degenerative disorders or head trauma.

As this example shows, it can be difficult to recognize early tics and associated behavioural problems outside the neuropaediatric speciality. Patients who may need help can remain undiagnosed as a consequence. Parents often tend to attribute the tics to psychological or local causes (e.g. conjunctivitis when the child is blinking). In this case, the parent's primary concern was the pain apparently associated with their child's teeth [4].

The diagnosis was made only after searching for other causes of pain, even though the child herself quite clearly described her generalized obsessive–compulsive problem from the start. It was felt to be obligatory to provide treatment to ease the pain and also to exclude any physical disease associated with the symptoms before referral so that a definitive diagnosis could be made.

The tics were painful because they resulted in teeth grinding causing pain to the attachment of the teeth. The masticator muscles and the temporomandibular joints were also overloaded with facial pain as a result [11,13].

Although these cases are uncommon, some gastrointestinal inflammatory diseases may present with symptoms such as pain and reddish swellings in the mouth [14].

Local infections in the parodontium and jawbone may also cause intensive, prolonged pain with reddish swellings, but this girl had received antibiotics covering *Staphylococcus* and Gram-negative anaerobic *Streptococcus* which had little effect on her symptoms [15]. Tumours must always be suspected when there is unexplained tooth pain, tooth mobility, jaw swelling or numbness without any other cause [15,16].

Molar incisal hypomineralization and amelogenesis imperfecta are mineralization diseases which may cause hypersensitivity of the teeth [17,18]. They differ from the mineralization disease caused by vitamin-D-resistant rickets in that they often are possible to diagnose clinically while the latter condition needs histological analyses [19]. Vitamin-D-resistant rickets is a rare metabolic disease that causes hypomineralization, microbial invasion of pulpal tissues, and thereby, severe toothache that usually starts in the mandibular anterior teeth. For this patient, histopathological examination of a tooth excluded disorders of mineralization.

The girl had no tics at the examination, but could explain the compulsion and she was able to reproduce the movement with her jaw. This is in accordance with earlier findings [8] that patients with TS have

partial control of their tics at least for short periods of time. This is a useful clue to recognize tic disorders.

The DSM-IV criteria separates TS from other forms of tics, although it is generally accepted that tic disorders form a continuum from the simple transient forms to fully fledged TS. The onset is usually during late childhood or adolescence, and in about 66% of cases, facial tics are the first symptoms. The course is fluctuating, with a tendency to improvement with increasing age. Facial, neck and shoulder muscles are especially affected with movement or tics [4]. Several reports have indicated that self-destructive behaviours occur in one-quarter to half of all cases [20]. The tics which this girl shows have changed to some extent, but they have always affected her face and jaws. Her tics have a strong presence of compulsivity, and because of being so intense, they are more or less painful and destructive to the involved tissues. Noncontingent reinforcement therapy, an occlusal appliance and relaxation therapy is what can be done in the dental office to help her to minimize her pain, the wear and pressure on her teeth, and the tension in her jaw muscles [21].

The pathogenesis of tic disorders involves both genetic and environmental factors, and a number of theories have been proposed. Treatment is usually not required for simple motor tics, but phonic tics, especially coprolalia, may be a social handicap and may be treated effectively with a number of drugs.

The oral signs and symptoms associated with TS were reported in the original description by Gilles de la Tourette in 1885 [5]. The true prevalence of oral manifestations is unknown, but many destructive oral lesions have been reported, including dysfunction of the temporomandibular joint, and chronic lip and mucosal ulcers. Tooth extractions are one complication caused by chronic oral tics, and these have been reported in at least three other cases [7]. Indeed, in a review by Friedlander *et al*., it was stated that 'dentists must be familiar with both the florid and subtle oral pathologic changes known to be associated with the disorder' [6].

To the author's knowledge, this is the first case described whose disease led her to present first to a dental service provider.

It may be concluded from this report that good cooperation between dentists and child neurologists regarding patients with TS should be advocated to allow early diagnosis and prompt treatment, since a delay may prolong suffering.

What this case report adds

- This report describes the case of a child with Tourette syndrome in a girl who presented first to a dentist with oral self injurious behaviour.
- Differential diagnosis proved difficult and a number of oral and systemic disorders were considered and excluded before the diagnosis was made.

Why this case report is important for paediatric dentists

- Oral self injurious behaviour in children is likely to present first to a paediatric dentist and may be associated with facial tics.
- Differential diagnosis may need to exclude Tourette syndrome, especially when signs of other oral or systemic disturbances are lacking and there is evidence of compulsive and or hyperactive behaviour.
- Good referral pathways and co-operative working with paediatric specialists can help to facilitate early diagnosis and prompt care for children with this condition.

Résumé. Le comportement d'auto-mutilation (SIB) est une blessure délibérée du corps sans intention suicidaire et survient chez certains patients à pathologie psychiatrique, comportementale et du développement. Ce cas décrit une jeune fille de 4 ans avec SIB, chez un spécialiste de dentisterie pédiatrique, provoquant une auto-extraction des dents, suite à des tics moteurs. L'enfant mobilisait régulièrement ses dents par un mouvement monophasique latéral entraînant une luxation de la canine temporaire maxillaire droite et provoquant des douleurs faciales et buccales généralisées. Les parents ont consulté le dentiste car leur enfant se plaignait de maux de dents. L'état buccal était tout à fait convenable à l'exception d'une canine temporaire mobile, mais il y avait des antécédents de comportement inhabituel incluant hyperactivité et après consultation multidisciplinaire et exclusion d'une autre maladie systémique, la patiente a été diagnostiquée comme souffrant du syndrome de Gilles de la Tourette (TS).

Un traitement préventif par contention a été réalisé. Une thérapie de renforcement non contingente a été appliquée avec succès pour diminuer le SIB.

Zusammenfassung. Selbstverletzung (SV) als eine intendierte Handlung mit schädlicher physischer Auswirkung auf den Körper ohne zugrundeliegende Suizidalität tritt bei einer Reihe von psychiatrischen Erkrankungen, Verhaltensstörungen und Entwicklungsstörungen auf.

Dieser Fall beschreibt ein 4jähriges Mädchen mit SV, das dem Zahnarzt mit Selbstextraktion von Zähnen infolge des Habits vorgestellt wurde. Das Mädchen

knirschte wiederholt in einer einseitigen Laterotrusionsbewegung, welche ihren rechten Oberkiefer Milcheckzahn luxierte und generalisierte Mund- und Gesichtsschmerzen verursachte. Die Eltern konsultierten einen Zahnarzt bezüglich der Beschwerden des Kindes. Die zahnärztlichen Befunde waren nicht richtungsweisend außer einer Lockerung des Milcheckzahns. Darüber hinaus lagen jedoch Verhaltensauffälligkeiten einschließlich Hyperaktivität vor. Nach einer multidisziplinären Abklärung und Ausschluss anderer systemischer Erkrankungen wurde die Diagnose eines Tourette Syndroms gestellt. Eine präventive Schienung wurde durchgeführt. Eine verhaltenstherapeutische Behandlung wurde erfolgreich zur Reduzierung des SV eingesetzt.

Resumen. La conduta de auto-lesiones (CAL) es un daño deliberado al cuerpo sin intento de suicidio y ocurre en varias alteraciones psiquiátricas, de conducta y del desarrollo.

Este caso describe una niña de 4 años que se presentó al odontopediatra con CAL produciendo la auto-extracción de los dientes como resultado de tics motores de la boca. La niña mordía repetidamente sus dientes en un movimiento lateral monofásico que producía la luxación del canino superior derecho resultando dolor bucal y facial generalizado.

Los padres consulatron al dentista por la queja de dolor del niño.

Las características bucales eran impecables exceto la movilidad del canino primario pero había una historia comportamiento inusual que incluía hiperactividad y después de consultas multidisciplinarias y de exclusión de otra enfermedad sistémica, se diagnosticó al paciente de enfermedad de Gilles de la Tourette.

Se realizó tratamiento preventivo usando una férula dental. Para disminuir el CAL se realizó con éxito la terapia de refuerzo no contingente.

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