Case Report

Clinical Presentation and Management of Peripheral-induced Oromandibular Dystonia in Nigeria: A Case Report and Literature Update

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Abstract

Oromandibular dystonia (OMD) is a rare focal neurological disorder associated with impaired masticatory function, dysphagia, dysphonia, and involuntary abnormal movements of the mandible of varying severity. The peripheral-induced variant among other factors is a common aetiological factor of secondary dystonias, associated with constellations of clinical features and presents with differing responses to various treatment therapies. To report a case of oromandibular dystonia and to add to the body of literature. The case report is of a 33-year-old male who presented with complaints pain on speaking and from the temporomandibular joint area for 8 months which adversely affected patient's work, social life and psychological well-being. Examination revealed retraction and repetitive but uncoordinated pattern of labial, cheek and masticatory muscular movement on mouth opening and closing. Diagnosis of oromandibular dystonia was made on clinical grounds. Patient was managed with a combination of medications by the oral physician, and physical therapy, which led to a significant improvement in the control of muscle contractions within 5 weeks. OMD though a rare neuromuscular condition can present to the dentist, and effective assessment, diagnosis and collaboration with relevant specialities are key to improved outcome.

Keywords: Botulinum toxin, dystonia, focal dystonia, oromandibular dystonia

Introduction

The term dystonia is used to describe a syndrome characterized by prolonged muscle contractions causing sustained twisting movements and abnormal postures of the affected body part(s).[1] Oromandibular dystonia (OMD) is a focal dystonia described as a neurological disorder that affects the mouth, face, and jaws.[2,3] It is defined as a movement disorder characterised by involuntary, paroxysmal, and patterned muscle contractions of varying severity, resulting in sustained spasms of masticatory muscles, affecting the jaws, tongue, face, and pharynx.[4] OMD can be classified as jaw opening, jaw closing, jaw deviating, or lingual dystonia or any combination of these.[5-7] When OMD coexists with blepharospasm and dystonic movements of the upper face, it is called Meige's syndrome.[2,8] OMD has a gender predilection with a female-to-male ratio of 3:1. The reported prevalence rates vary widely from 50 cases/million for early-onset primary dystonia and 30–7320 cases per million for late-onset primary dystonia.[9] The overall prevalence of cranial and focal dystonia is not clearly defined yet; however, a regional U. S. prevalence study estimated 69/million cases for OMD[9] and the incidence is projected to be up to 3.3 cases/million.[10] There were no available local or regional data on the prevalence of OMD at the time of this report. However, Blunt *et al*. reported a case of orolingual dystonia in a 54-year-old woman of Nigerian descent.[11]

The onset of symptoms is usually between the ages of 40 and 70 years.[3] The symptoms only occur during activities such

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as speaking or mastication[3] and clinical presentations depend on the affected muscle, as well the severity and distribution of OMD.[12] Clinical manifestations include impaired mastication, dysphagia, dysphonia, unconscious opening and closing of the mandible, pulling and twisting of the mandible forward or laterally, and temporomandibular disorders such as open locks.[7,13] Tongue protrusion is also a component of OMD or Meige's syndrome.[14]

While the genetic basis for primary dystonia and recognisable disorders for secondary dystonia have been proposed, the mechanism and cause of the development of OMD have not been fully elucidated.[3,15] OMD may occur as a spontaneous neurological disorder, with or without a hereditary history, or attributed secondarily to certain drugs or disorders such as trauma or Wilson's disease.[3] Iatrogenic trauma from dental treatment has also gained traction as a recognisable etiological factor.[16]

Diagnosis of OMD is based on the clinical history and neurological examinations; however, specific investigations may be carried out to confirm any of the etiological factors. Raoofi has suggested that intramuscular electromyography can be used as a confirmatory test.[3] Treatment of OMD is multidisciplinary and personalised for every patient. Modalities include medication, physiotherapy, botulinum toxin (BoNT) injections, muscle afferent block (MAB), surgical therapies, and other therapeutic approaches, including cognitive behavioral therapy.[3]

This case report aims to report findings and draw attention to this rare neuromuscular condition "OMD" which was presented at the oral medicine clinic of the University of Benin Teaching Hospital.

Case Report

A case of 33-year-old male patient was referred to the Oral Medicine Outpatient Clinic of the University of Benin Teaching Hospital on account of suspected temporomandibular joint (TMJ) disorder and dyskinesia. The patient complained of inability to talk properly, pain in speaking, and pain around the TMJ area, all of eight-month duration. There was also a history of uncontrollable and irregular movements of the tongue while speaking. He also reported occasional unilateral retraction of the lip, cheek, and jaw on attempts to speak. These movements were often spontaneous but mainly triggered by speaking, eating or chewing, yawning, smiling, or laughing. Movements are usually continuous and sustained until the patient decides to relax. There was mild-to-moderate pain that accompanied movements of the jaw and radiated to the TMJ region. There was a positive history of the right TMJ traumatic dislocation following an assault four months before the onset of symptoms, and was reduced after seven months using the bimanual maneuver by a general dentist. There was no history of long-term use of antipsychotics, antidepressants, or anticonvulsants. There was also no history of head injury. The collection of symptoms has progressed and worsened over time. The patient is a married man with a child in a monogamous setting and a businessman. There is no history of a similar condition in his family. The patient also reported that the condition has adversely affected his work, social life, and psychological well-being.

On examination, the patient was calm and in no obvious distress, concealing and cleaning his mouth with a handkerchief. There was facial symmetry at rest, mouth opening of 5 mm interincisal distance. There was retraction and repetitive but uncoordinated pattern of labial, cheek, and masticatory muscular movement on mouth opening and closing, with a deviation of the mandible to the left on mouth opening and on an attempt at talking or laughing. Refer to Figures 1 and 2 for clinical images. No crepitus, tenderness, or clicking sound was detected on the TMJ. The tongue also retracts and deviates to the left or right on mouth opening and closing as shown in a short video 1 with a link published hereto. The cranial nerves examination was unremarkable.

A clinical impression of OMD was made. Open- and closed-mouth transcranial plain radiographs [Figures 3 and 4] were done to rule out temporomandibular disorder and revealed unequal forward excursions of the heads of the condyle from its joint socket. Their movements produced a shift of 3 cm on the right and 2 cm only on the left. This finding corroborated the marked restriction of movement on the left TMJ compared to the right. There was no sign of a permanent dislocation of either joint detectable on the radiograph. After clinical and radiologic evaluation, a diagnosis of OMD was sustained.

The patient was counselled and reassured of the controllability of the disease and the possibility of spontaneous remission. Diazepam 10 mg nocté for two weeks and ibuprofen 400 mg twice daily for three days were prescribed. At the two week follow up visit, there was little or no improvement, and the patient was commenced on Norgesic (orphenadrine citrate 25 mg, aspirin 385 mg, and caffeine 30 mg) two tablets twice daily for two weeks and oral Neurobion (Vitamins B1 [thiamine disulfide] 100 mg, Vitamin B6 [pyridoxine hydrochloride] 200 mg, and Vitamin B12 [cyanocobalamin] 200 µg) one tablet daily for one week. Also, topical Olfen (diclofenac) gel was recommended, to be applied on areas of pain and tenderness twice daily and regular jaw exercise by chewing sugarless gum.

There was a recorded improvement in the symptoms after another two weeks, and while the current medication continued, the patient was referred to physiotherapy for the treatment of muscle stiffness. There was a significant improvement on the third review three weeks later. Subsequently, diazepam was substituted with clonazepam 0.5 mg daily, and other medications and physiotherapy were continued. Although the patient has not been compliant with his review appointments, phone contact with the patient revealed an improving condition.

Discussion

The term "dystonia" is used to describe a syndrome characterised by prolonged muscle contractions causing sustained twisting movements and abnormal postures of the affected body part(s).[1] OMD, a focal dystonia, is a rare clinical entity with reported female preponderance[9,17] and average age of onset of 51.6 years.[17] This report, to the best of our knowledge, is the first report in national literature which supports its rareness.

The etiology of OMD may be idiopathic or secondary. The main mechanism of onset of OMD is largely unknown; however, some pathophysiological explanations have been suggested as likely causes, such as basal ganglia dysfunction, hyperexcitability of motor neurons interneurons related to signaling pathways, loss of inhibition, aberrant dopamine signaling, monoaminergic dysfunction, abnormal plasticity, and abnormal sensory function.[18,19] Idiopathic dystonia attributed to a presumed or actual genetic cause account for the majority (>60%) of cases.[20]

Secondary dystonias are usually a result of peripheral trauma or surgical incidents; neurodegenerative disorders; cerebral infarction; and use of drugs (tardive dystonia) such as neuroleptics, antiemetics, antimalarial, carbamazepine, and cephalosporins.[20-22] Other less frequent etiologic factors of secondary OMD are postanoxic states in 2.5%, neurodegenerative disorders in 1.8%, and head injury-associated OMD in 0.9%.[23,24]

Bakke *et al*. reported 21 cases of OMD, 2 of which were related to injury or infection.[7] A case of dental implant-induced OMD has also been reported.[25] The reported case has a positive history of trauma to the jaw region, which was poorly managed.

A number of genes known as DYT genes have been associated with etiopathogenesis of OMD; however, the exact mechanism is still largely sketchy.[15] They include proteins that appear to function as chaperones (DYT1), transcription factors (DYT6), structural proteins (DYT11), and enzymes involved in dopamine biosynthesis (DYT5).[15] For instance, mutation in the DYT1 gene could lead to young-onset primary dystonia in children and adolescents.

Clinical presentation of repetitive or sustained involuntary prolonged spastic movements of the tongue, facial, masticator, and perioral muscles are very essential in making a diagnosis. Our patient presented with dystonic movements of the perioral, masticatory, lower facial muscle, and the tongue without involvement of the upper face and periorbital structures. The symptoms only occur during activities such as speaking and mastication as observed in this case.[3] OMD can be classified as jaw opening, jaw closing, jaw deviating, or lingual dystonia or a combination of these,[5,7,12] as seen in the reported case. Even though the index case presented with pain during jaw movements around the area of the TMJ, pain has been a rarely reported feature.[3]

The severity of symptoms and progression of the disease are more prominent in posttraumatic OMD than in primary OMD.[3] Family history of movement disorders is also less frequent in posttraumatic OMD,[20] as was absent in this case.

Figure 1: Clinical photograph showing a sustained pattern of labial and cheek muscular contractions

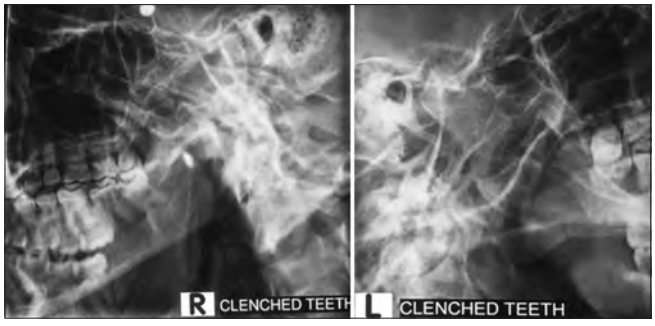

Figure 3: Right and left clenched teeth transcranial view radiograph

Figure 2: Clinical photograph showing a sustained pattern of labial and jaw muscular contractions

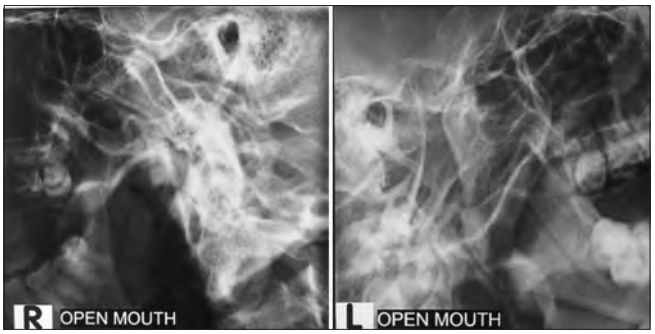

Figure 4: Right and Left open mouth transcranial view radiograph showing the unequal excursion of the condyles on the open mouth position

OMD may sometimes coexist with other syndromes/diseases such as Wilson disease, Huntington disease, Niemann–Pick disease, Leigh disease, and Parkinson's syndrome.[26]

The diagnosis of OMD is clinical,[27] based on history and physical and neurological examination. There is no test to confirm the diagnosis of OMD, and in most cases, routine laboratory tests are normal.[7] In most cases, the oral dysfunction is associated with social embarrassment, reduced quality of life, and depression.[27] In this case, a history of trauma and subsequent dislocation of TMJ ruled out differential diagnoses such as, bruxism, spontaneous condylar dislocation, facial spasm, and psychological disorders.[28]

OMD can lead to afflicting outcomes that adversely affect the day-to-day living of affected persons, like in the index case. OMD has been associated to a reduction in the quality of life and depression.[7] Esthetics alterations from jaw deviation or involuntary tongue protrusions can also elicit feelings of embarrassment and diminished self-confidence.[5,29] Severe cases may lead to difficulty with swallowing or speech and dental trauma.[30] The index case presented similarly with dysphagia, pain, and difficulty in chewing; however, there were no signs of dental trauma.

Although treatment of OMD should be multidisciplinary, it should be tailored to individual patient needs. The aim of the treatment is to reduce neuromotor signal discharge and achieve motor stability, to inhibit muscular excitability and prevent excessive muscle contraction, to relieve symptom and pain, and to improve quality of life. Currently, there is no curative treatment for OMD, but several treatment strategies may help to reduce the severity of the symptoms.[31] These strategies include botulinum toxin (BoNT) injections, pharmacological treatment, physical and behavioral therapy, deep brain stimulation, surgical therapy, MAB, and others include psychosocial and occupational therapy, support group participation.[4] Similarly, the index case was treated with medications and jaw exercise as a physical therapy. This combination therapy showed considerable effectiveness as our patient experienced relief of symptoms, better muscular control, and improved quality of life within three weeks of therapy. BoNT injections have particularly gained traction as the future prospect, baring the fact that BoNT requires trained expertise for safe and effective application. For instance, a study carried out in a dystonia coalition international multicenter involving 727 OMD subjects, BoNT provided symptomatic relief by more than 50% irrespective of the etiology in about 80% of the subjects.[27] Ameer and Bhatti 2021 shared their BoNT injections experience as the best treatment modality, offering long-term relief with lesser side effects.[32] However, there are still gaps to fill, perhaps a need for large randomised clinical trials.

Conclusion

OMD, though a rare neuromuscular condition, can present to the dentist, and effective assessment diagnosis and collaboration with relevant specialties are key. The reported patient responded well to a combination of therapy with medications and physiotherapy in this study. Prompt diagnosis and early treatment intervention in the presented case formed the key to a successful outcome observed in this patient.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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