Masticatory Muscle Pain and Progressive Mouth Opening Limitation Caused by Amyotrophic Lateral Sclerosis: A Case Report

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This article reports a case of masticatory muscle pain and progressive limited mouth opening secondary to amyotrophic lateral sclerosis (ALS), popularly known as Lou Gehrig's disease. The symptoms were first mistaken as those of temporomandibular disorders, before fatty degeneration of all masticatory muscles were discovered on magnetic resonance imaging (MRI). ALS should be considered in the differential diagnosis process when the patient presents with longstanding progressive mouth opening limitation associated with pain. MRI could facilitate the diagnostic process. *J Oral Facial Pain Headache 2015;29:91–96. doi: 10.11607/ofph.1340*

Key words: *amyotrophic lateral sclerosis, motor neuron disease, mouth opening limitation, orofacial pain*

Amyotrophic lateral sclerosis (ALS), which is also known as Lou Gehrig's disease, is the most common adult-onset motor neuron disease. The disease shows selective and progressive degeneration of motor neurons, which results in spasticity, hyperexcitability, and appearance of pathologic reflexes caused by degeneration of upper motor neurons of the cerebral cortex, as well as muscle weakness and atrophy followed by progressive paralysis caused by the degeneration of lower motor neurons of the brainstem and anterior horn of the spinal cord.[1,2] Incidence rates are relatively low in the general population, being 1.5 to 2.5 per 1,000,000 persons per year.[3] The consequences of the progressive degeneration of motor neurons that innervate voluntary muscles are generally fatal, since respiratory failure usually eventuates and results in death. The average median survival is 19 months from diagnosis and 30 months from onset.[4] The etiology of ALS is yet to be elucidated, but accumulated evidence suggests that, similar to other chronic neurodegenerative diseases, ALS is a complex genetic disorder with different subtypes according to genetic mutations and their interaction with environmental risk factors.

In the following case, the patient was initially mistaken as presenting features indicative of temporomandibular disorders (TMD) due to the patient's masticatory muscle pain and limitations of mouth opening. However, the severity and duration of the limited mouth opening raised the need for further imaging.

Case Report

A 48-year-old male patient visited the Orofacial Pain Clinic, Department of Oral Medicine and Oral Diagnosis, Seoul National University Dental Hospital with the chief complaint of stiffness of both masseter muscles during function and progressive limitations of mouth opening that recently seemed to be getting worse. The decreased mouth opening range had first been noticed 5 years earlier. The patient reported a history of open lock after maximum mouth opening and that diffuse pain had appeared in the shoulder, low back, and thigh area. Such systemic pain had recently progressed to have an intermittent electrical aspect.

Table 1 RDC/TMD Assessment and Imaging Results of the Case

	Score/Result
Axis II	
Characteristic pain intensity	60 (0–100)*
Graded Chronic Pain Scale	Grade 2 (1–4)*
Depressive and vegetative symptoms	1.889 (0–4)*
Somatization	1.9 (0–4)*
Jaw Functional Limitation Scale	0.5 (0–1)*
Imaging	
Plain radiograph	DJD of both TMJ condyles
Computed tomography	DJD of both TMJ condyles
Magnetic resonance imaging	1) ADD with reduction of right TMJ condyle 2) Fatty degeneration of all masticatory muscles of both sides

*Range of score.
DJD, degenerative joint disease; TMJ = temporomandibular joint; ADD = anterior disc displacement.

The patient first visited the Department of Neurology in a tertiary care hospital for examination. When further asked about the results, he incorrectly reported that the test results were within normal range and a specific diagnosis was not given. He reported that the recent weather with low temperatures had been aggravating the symptoms for the past 2 to 3 months. Past medical history was not noteworthy, except for a tuberculosis infection 20 years ago. He was currently taking over-the-counter analgesics on a pro re nata basis when the pain was severe enough to interfere with daily activities.

Intraoral and Extraoral Findings

Findings from the intraoral examination were unremarkable. Routine laboratory blood tests including complete blood cell count with differential and erythrocyte sedimentation rate showed normal results.

Extraoral examinations were conducted following the Research Diagnostic Criteria for Temporomandibular Disorders (RDC/TMD).[5] The patient reported pain on palpation of the posterior portion of both temporalis, superior portion of both sternocleidomastoid, and both trapezius muscles. The maximum mouth opening range was 20 mm. His chin deviated to the left on mouth opening. No joint noises could be observed. A check of the occlusion with the use of an 8-µm-thick film showed a sound occlusion. The patient reported unilateral chewing, insomnia, indigestion, and an irregular diet pattern during the investigation for possible contributing factors.

The Symptom Check List-90-Revised[6] was applied for psychological evaluation, and the results were within normal range. The analysis of his answers on the RDC/TMD Axis II questionnaire revealed a characteristic pain intensity of 60 (on a 0–100 scale). The patient was classified as grade II (low disability, high intensity) on the Graded Chronic Pain Scale.[7] The scores for depressive and vegetative symptoms and somatization indicated the possibility of severe depression (≥ 1.105) and somatization (≥ 1.000). Jaw disability was assessed with the Jaw Functional Limitation Scale[8]; the patient expressed difficulty in chewing, tooth brushing, exercising, chewing hard food, swallowing, and talking using the scale's checklist. Axis II scores are shown in Table 1.

The patient was initially diagnosed as RDC/TMD group Ia, myofascial pain with limited opening, without knowing his correct medical history. The possibility of bony ankylosis of the temporomandibular joint (TMJ) or muscle contracture was also considered.

Imaging Findings

The severity and duration of the patient's limited mouth opening raised the need for further imaging. Plain radiographs including orthopantomogram, TMJ panoramic view, and transcranial view were evaluated. Computed tomography and magnetic resonance imaging (MRI) were also carried out. Results are shown in Figs 1 to 3. Osteophytic bone formation could be seen on the anterior portion of both TMJ condyles. The movements of the TMJ condyles were markedly restricted, with the condyles remaining inside the mandibular fossa during maximum mouth opening. MRI revealed advanced fatty atrophy with reduced volume of both sides of the masseter, medial pterygoid, lateral pterygoid, and temporalis muscles.

The possibility of a neurologic disorder or metabolic disorder was suggested to the patient along with the need for further investigations. Then the patient revealed his full medical history of being diagnosed with ALS in the past and not obtaining any further treatment specific to his condition from then on.

Medical Findings

Nerve conduction and electromyographic studies revealed denervation potentials in three compartments (cervical, thoracic, vulva) including the tongue. Mild atrophy of the tongue was also observed. Additional laboratory tests, including thyroid function tests and serum protein electrophoresis along with cervical spine MRI, did not show any pathologic findings. Tests for autoimmunity all showed negative results. Genetic testing showed that the CAG trinucleotide repeat number of the causative gene, the androgen receptor gene, was 25 (normal limit < 33 repeats). No tumor markers were identified. A videofluoroscopic study of swallowing revealed a severe vallecular fossa and pyriform collection of fluid when the patient swallowed in a chin-up position. Tracheal penetration and aspiration were also seen during bolus swallowing in the

chin-up position. Cough reflex was not appropriately present at the time that the diagnosis of ALS (progressive muscle atrophy variant) was made, and Levin tube feeding was recommended along with rehabilitation therapy for dysphagia. The patient repeatedly reported difficulty in swallowing and chewing.

Treatment

Gabapentin in combination with the nonsteroidal anti-inflammatory drug (NSAID) aceclofenac was initially prescribed by the Department of Neurology to control the pain, but this was subsequently replaced by a synthetic opioid (tramadol) for more powerful pain relief.

On the first visit to the orofacial pain clinic, instructions for control of habits that may aggravate the masticatory muscle pain were given. After the MRI results and the patient's true medical history were reviewed, physical therapy instructions including moist hot pack application, progressive resistance exercises, and Rocabado's 6 × 6 exercise were given. However, the patient stated that his neurologist had directed him to avoid exercising. Additional NSAIDs (aceclofenac) were prescribed for pain control.

Discussion

This is the first case report to describe the management of a patient with ALS who presented with limitations of mouth opening and masticatory muscle pain as his chief complaints. The patient was mistakenly diagnosed as having myofascial pain with limited mouth opening that reflected RDC/TMD group Ia, based only on clinical examinations and plain radiographs.

Setting aside the false medical history report by the patient, this case reveals that the clinician who manages orofacial pain must be well acquainted with the signs and symptoms of ALS, since pain is a frequently accompanying symptom. Previous studies have reported that pain usually of a dull or electrical quality occurs in up to 80% of the cases.[9] Some patients may report various types of pain in multiple parts of the body. The severity of pain is generally mild to moderate, with 20% of patients reporting severe levels of pain.[10,11] In the initial stage of the disease, symptoms due to neuron damage and resultant muscle atrophy may be less evident, and pain solely appearing in the orofacial region may easily be misdiagnosed as TMD. To avoid such errors having grave consequences, the clinician must be sufficiently informed of the early signs and symptoms of ALS, which commonly start in the limbs and include foot drop, difficulty in walking, weakness when lifting the arms, and loss of hand dexterity. Symptoms that may occur in the orofacial region are dysarthria followed by dysphagia, and sialorrhea may appear as the disease progresses.[12] However, in the present case, the patient's pronunciation and gait appeared to be normal. The patient answered "yes" to difficulty in chewing, tooth brushing, exercising, chewing hard food, swallowing, and talking from the Jaw

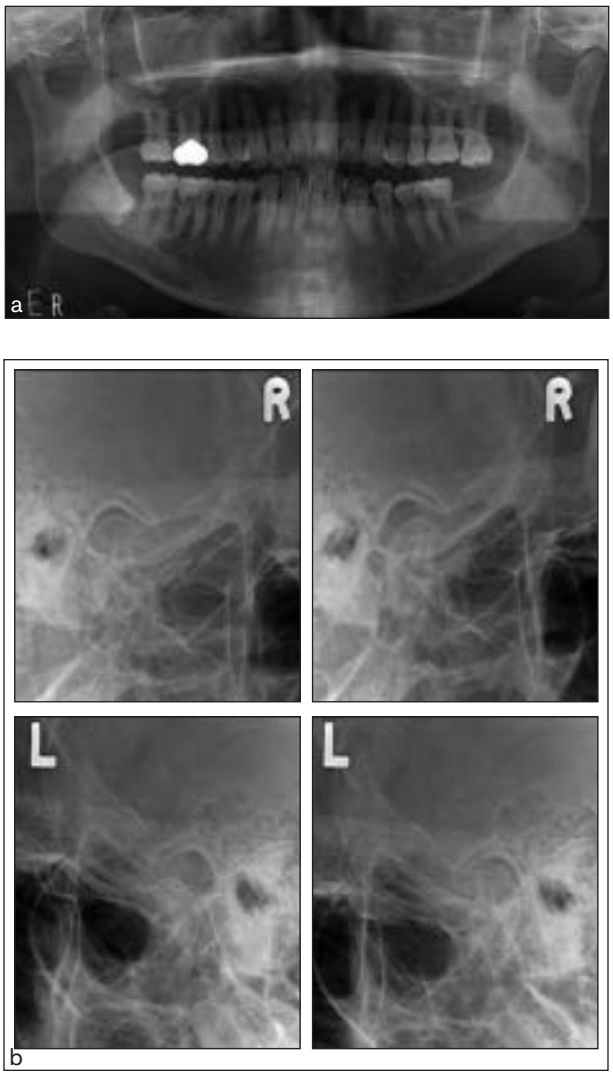

Fig 1 Plain radiographs of the TMJs. **(a)** Orthopantomogram showing degenerative change of both TMJ condyles. **(b)** Tr ans–cranial view of the TMJ showing marked restriction of condylar movement with the condyles remaining inside the mandibular fossa during maximum mouth opening.

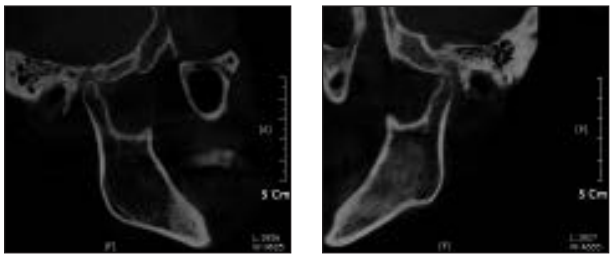

Fig 2 Computed tomography of the TMJs. Sagittal view of the TMJs showing slight osteophyte formation on both TMJ condyles.

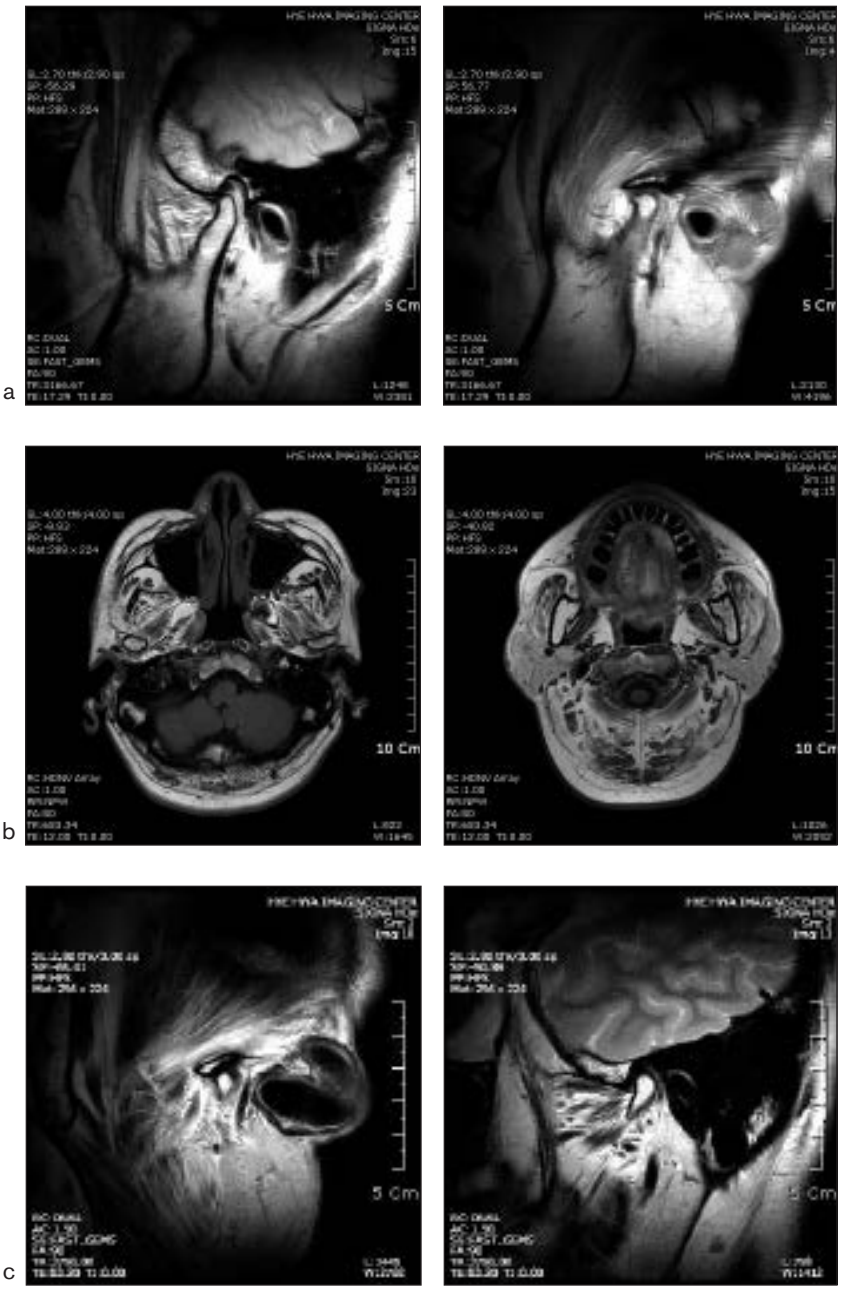

Fig 3 MRI of the TMJs. **(a)** T2-weighted sagittal MRI showing slight anterior disc displacement at closed-mouth and reduction of the disc at open-mouth position of the right TMJ. The movements of both TMJ condyles show severe restriction at maximum mouth opening. The condyles are showing beaking at the anterior portion (left side: closed mouth; right side: open mouth). **(b)** T2-weighted axial MRI showing advanced fatty degeneration of the masticatory muscles (left side: lateral pterygoid muscle; right side: medial pterygoid and masseter muscle). **(c)** T2-weighted sagittal MRI showing advanced fatty degeneration of the temporal, masseter, and lateral pterygoid muscles (left side: left temporal and masseter muscles; right side: left lateral pterygoid muscle).

Functional Limitation Scale checklist. Additional questions about episodes of sialorrhea or food spilling from the mouth may have more easily raised the clinician's suspicions of a possible motor neuron disease.

Dysphagia and pain of the masticatory muscles may result in malnutrition. Frequent evaluations of the nutritional status and management must be made in view of the common occurrence of dysphagia.[13] Although dentists may not carry out full-scale nutritional assessments, the possibility of malnutrition should always be considered when treating an ALS patient, and vitamin and mineral supplements should be recommended, since they are known to prolong the survival of ALS patients.[14]

Cognitive impairment, which appears in about 50% of ALS patients, is another aspect of ALS that requires attention.[15,16] Such impairment may lead to communication difficulties that further increase the possibility of misdiagnosis. Verbal fluency is known as a sensitive index of cognitive impairment level in ALS that may be used to differentiate patients.[17] Another confounder of the diagnostic process is the depression and anxiety that can occur throughout the progression of ALS.[18] Depression and anxiety

are also well-known features of TMD pain.[19,20] Such psychological symptoms may have secondary effects on the level of orofacial pain through lowered sleep quality and catastrophizing.

Pain in ALS is generally treated with non-opioid analgesics such as NSAIDs, as in the present case.[21] However, there is no clear consensus on the dosage, duration, and method of administration of such analgesics. The clinician must consider the possibility of drug interactions when prescribing additional drugs and more actively participate in controlling pain, especially when the major location of pain is the orofacial region. Uncontrolled pain in the ALS population definitely lowers the patient's quality of life, yet pain is frequently undermanaged.[22] The majority of patients with ALS are known to take riluzole, which reduces excitotoxicity, so the clinician must be aware of possible side effects and drug interactions.[23] Common side effects include diarrhea, dizziness, fatigue, nausea, and somnolence.

The muscle pain in ALS patients may originate from muscle contractures and joint stiffness.[24] This may explain why physical therapy and therapeutic exercises are reported to reduce the pain and increase the quality of life of ALS patients.[25,26] Many physicians still do not recognize the benefits of physical therapy and exercises, as in the present case, and direct the patient to avoid any forms of exercise based on the possibility of further burdening the weakened musculature. However, considering the etiology of pain in ALS patients, appropriate exercises may reduce muscle and joint stiffness. Research is lacking in establishing the method and intensity of exercises and physical therapy for ALS patients, and no recommendation exists for the masticatory muscles. Moist hot-pack application may be considered along with flexibility and isometric exercises. Passive stretching that does not aggravate pain may help maintain an acceptable mouth opening range. Since no guideline exists, further studies are necessary to determine the specific loading and duration for exercises and physical therapy that will not cause additional fatigue for the masticatory muscles.

Reports of botulinum toxin injection for excessive salivation in ALS patients reveal that dental experts may play a more active role in the treatment of ALS patients.[27] Since there is no cure for ALS, the objective of clinical care is focused on maintaining the patient's quality of life and extending the patient's survival. A multidisciplinary approach is needed, and the dentist should actively participate as a member of the health care team in controlling symptoms such as pain of the orofacial region and sialorrhea.

In addition, the importance of proper imaging cannot be overemphasized in the diagnostic process for patients with orofacial pain due to ALS. The present case was diagnosed as TMD based on only clinical examinations, laboratory tests, and plain radiographs. It was possible to raise doubt on this initial diagnosis only after the results of the MRI showed fatty atrophy of all masticatory muscles. The current RDC/TMD methodology shows high reliability and reproducibility but fails to place emphasis on additional imaging tests. A high rate of false results have been reported when solely depending on RDC/TMD clinical examinations.[28] Additional MRI should be considered when the patient presents with chronic and progressive limitations of mouth opening without improvement.

Conclusions

ALS can be associated with chronic orofacial pain that may lead to a misdiagnosis of the condition. The dental expert should be well informed of the signs and symptoms of ALS to avoid such errors. MRI can facilitate the diagnostic process when a patient presents with progressive long-standing limitations of mouth opening accompanied by masticatory muscle pain.

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