Clinical Reasoning:
A case of unilateral facial pain

SECTION 1: CLINICAL PRESENTATION
A 62-year-old man admitted with B cell chronic lymphocytic leukemia (CLL) reported 2 weeks of progressive left-sided jaw and facial pain. He first noticed the pain occurring during prolonged chewing. It was exacerbated by teeth clenching and resolved with jaw rest. The patient described the pain as dull and gnawing without radiation. He also described a “knot” within his left “jaw muscles” at the mandibular angle. He had decreased range of motion, could not fully open his mouth, and felt that his jaw was misaligned. He was therefore placed on a soft mechanical diet. He denied a temporal headache or transient loss of vision. He was on his second day of obinutuzumab chemotherapy but was not on any other new medications. He had no history of trauma to his face, head, or neck.

The most notable abnormalities on examination were that his left masseter and temporalis muscles were more prominent than the right, both at rest and when clenching. His left masseter was tender, and his jaw’s range of motion was reduced and limited by pain. On general head and neck examination, there was no temporal artery tortuosity. There was no tenderness of either of the temporomandibular joints (TMJs) and no parotid gland swelling. There was no lateral mandibular deviation or clicking sounds from the TMJ, and there was no neck dystonia. Oral examination did not reveal abscesses or other signs of inflammation.

Cranial nerve examination revealed normal visual acuity, visual fields, and extraocular movements. The patient had normal perception of pain, light touch, and temperature sensation in V1-V3 bilaterally, without hyperesthesia or allodynia. Lateral pterygoid function was preserved. Frontalis activation, eye closure, lip pursing, and smile were symmetrical and normal. Hearing was normal. Tongue and pharyngeal muscle movements were normal. The remainder of the neurologic examination revealed no other pathologic or lateralizing features.

Questions to consider:
1. What is the localization of the lesion?
2. What is the differential diagnosis?
SECTION 2: LOCALIZATION AND DIFFERENTIAL DIAGNOSIS

Localization. The localization most likely involves a lesion of the peripheral nervous system or masticatory muscle. The isolated masseter muscle tenderness and lack of sensory symptoms or other cranial nerve findings make spinal trigeminal tract, brainstem, and thalamic lesions much less likely. The masticatory muscle asymmetry could represent either left muscle hypertrophy or right muscle atrophy. While a lesion of the right trigeminal motor nucleus could cause right-sided atrophy, cortico-trigeminal tracts project bilaterally, making this sort of lesion unlikely. However, ischemic or compressive damage to the motor fibers of the mandibular division of the trigeminal nerve as it exits from the skull could cause right-sided atrophy. All together, the clinical description supports a peripheral process, comprising the masticatory muscles and the surrounding soft tissue, with possible involvement of motor components of the right trigeminal nerve.

Differential diagnosis. The asymmetric tender masseter muscle suggests a diagnosis of masticatory myalgia as opposed to temporomandibular joint dysfunction, vascular claudication, glandular disease, or systemic inflammatory disease. The differential diagnosis includes compressive lesions of the mandibular division of the trigeminal nerve or diseases of the muscle such as neoplasia of the masticatory muscles, masseter muscle myopathy, or idiopathic masseter muscle hypertrophy. Systemic inflammatory diseases such as giant cell arteritis, sarcoidosis, and Sjögren syndrome, temporomandibular joint disorders, or parotid gland diseases are lower on the differential because of the lack of corroborating clinical signs on examination, but still a consideration because of the pain with prolonged mastication and reduced jaw opening.\(^1\)\(^-\)\(^3\) Of these diagnoses, the history of CLL raised the suspicion for metastasis to either the left masticatory muscles or the right motor trigeminal nerve.

Questions to consider:

1. What laboratory investigations might help narrow the differential diagnosis further?
2. What if any neuroimaging would you obtain?
SECTION 3: INVESTIGATIONS
Several laboratory investigations could be considered based on the pretest probability of each differential diagnosis. In this case, giant cell arteritis was strongly considered. However, the erythrocyte sedimentation rate and C-reactive protein level were within the normal range. Anti-Ro and anti-La antibodies were not detected, making Sjögren syndrome a less likely candidate.

MRI of the brain, face, and neck, with and without contrast, confirmed asymmetry of the masseter and temporalis muscles (figure 1) and revealed a right infratemporal fossa lobulated cystic lesion near the foramen ovale (figure 2). There was left-sided TMJ osteoarthritis with mild synovitis as well (not shown). There was no radiographic evidence of intramuscular lesions to suggest tumor, hemangioma, or inflammatory changes.

Question to consider:
1. What is your final diagnosis?

Figure 1  Axial MRI of the masticatory muscles

Axial T1-weighted MRI demonstrates right masseter (A) and temporalis muscle (B) atrophy with left muscle hypertrophy (arrowhead points to left masseter muscle, arrow points to left temporalis muscle).

Figure 2  Coronal MRI demonstrates foramen ovale cyst

Coronal MRI demonstrates a sharply contoured lobulated cystic lesion that appears hyperintense on T2 (A and B) and is non-enhancing and hypointense on T1 with contrast (C), located in the right infratemporal fossa involving the foramen ovale and extending subtemporally.
SECTION 4: DIAGNOSIS AND DISCUSSION

The final diagnosis is a right infratemporal fossa lobulated cyst, causing compressive neuropathy of the V3 segment of the right trigeminal nerve as it exits the foramen ovale. The consequent right temporals and masseter muscle atrophy likely caused overcompensation of the left muscles of mastication and left (TMJ) osteoarthritis and inflammation. Although there can be several causes of facial pain that are not primarily neurologic, in this case, the left TMJ inflammation (typically a non-neurologic problem) is likely attributable to neuropathy of the contralateral trigeminal motor nerve. Despite evidence of right-sided masticatory muscle atrophy on MRI, the patient reported subacute onset left face pain. Denervation-related alterations in muscle bulk can occur along a spectrum from acute hypertrophic to chronic atrophic changes. The atrophy would seem to suggest a more chronic disease process occurring on the right. We suppose that the subacute on chronic presentation could be secondary to enlargement of the cystic lesion causing more dramatic changes in masticatory function, subacute compensatory hypertrophy involving the left masticatory muscles, or worsening inflammation of the left synovial joint tissue. It is unclear if the malignancy could have contributed to a systemic inflammatory response worsening the symptoms of compressive cranial neuropathy.

There are several important teaching points of this case. First, when evaluating facial pain, a detailed examination of the head, neck, and cranial nerves is necessary. Facial pain can be accompanied by visual disturbances, abnormal eye movements, pupillomotor dysfunction, or facial sensation and may be seen with facial palsy, hypoacusis, balance disturbance, dysphagia, dysphonia, or dysarthria. These signs, if present, assist in localization. Secondly, the motor component of the trigeminal nerve may be easily overlooked without change of facial sensation. The motor portion of the trigeminal nerve has its nucleus in the pontine tegmen tum. In the trigeminal cave (Meckel cave), the fascicular portion joins sensory fibers as it becomes incorporated into the mandibular nerve. The mandibular nerve exits the skull through the foramen ovale and supplies the masseter, temporalis, pterygoid, mylohyoid, tensor veli palatine, and the anterior belly of the digastric muscle. A lesion anywhere along this path can cause motor involvement. Additionally, selective motor involvement of the mandibular nerve has been described. third, it is useful to consider the contralateral consequences of unilateral loss of a paired cranial nerve. Finally, cranial neuropathies or asymmetry of cranial nerve function should raise suspicion for a lesion involving the foramina through which the cranial nerves exit the skull.

Unlike this case, false localizations are most commonly seen with increased intracranial pressure or spinal cord lesions. Increased intracranial pressure is known to cause apparent sixth cranial nerve palsy, but has also been reported to cause fifth and seventh cranial neuropathies, fifth cranial neuralgia, hemifacial spasm, third cranial neuropathy, and contralateral hemiparesis secondary to Kernohan notch phenomenon. Lesions at the level of the foramen magnum may produce upper motor neuron signs, but also signs of paresthesia in the hands and lower motor neuron signs in the upper limbs.

The role of imaging depends on localization and clinical suspicion. MRI is most specific and sensitive for soft tissue and inflammatory conditions of the joint, whereas x-ray and CT can demonstrate osseous morphology. MRI is suggested when evaluating cranial neuropathies, new onset neuralgias, or sudden-onset severe headache. In this case, we were able to clinically localize the lesion peripherally. However, the MRI was useful in further localizing the lesion to the foraminal segment of the trigeminal nerve.

TMJ disease is a common cause of jaw and facial pain and is characterized by pain, cracking or popping noises with jaw movement, and limitation of jaw movement. Patients require combined nonpharmacologic (education, behavioral modification, and occlusal splints) and pharmacologic treatment (nonsteroidal anti-inflammatory drugs [NSAIDs], muscle relaxants with NSAIDs, and tricyclic antidepressants). In extreme cases, surgical intervention may be necessary. In our patient, NSAIDs were contraindicated due to thrombocytopenia. The cyst was not amenable to surgical resection.

AUTHOR CONTRIBUTIONS

Both authors contributed to the conception, design, image acquisition, and writing of the manuscript and had final approval of the submitted manuscript.

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