

Misdiagnosis: Hypoglossal palsy mimicking bulbar onset amyotrophic lateral sclerosis

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ABSTRACT

Bulbar onset amyotrophic lateral sclerosis (bALS) is a currently incurable neurodegenerative condition characterized by insidious progression of bulbar muscle paresis; namely dyspnea, dysarthria, and dysphagia. The diagnosis of bALS requires exclusion of mimicking pathologies as the diagnosis of bALS may have significant implications on patients' quality of life, future planning, and familial/social dynamics. Herein we present two cases which were misdiagnosed as bALS when in fact a structural lesion of the hypoglossal nerve was causative. This article may serve as a reminder to entertain alternative diagnoses prior to arriving at a diagnosis of bALS.

1. Introduction

Bulbar onset amyotrophic lateral sclerosis (bALS) is a progressive neurodegenerative process involving both cerebral and spinal motor neurons with the first clinical signs appearing within the bulbar muscle groups [1,2]. Patients with bALS have a worse prognosis when compared to other phenotypes, making the diagnosis critically important for the patient and the patient's family [3]. Within this case series, we describe two cases which presented to our outpatient Neurology clinic for second opinion regarding a diagnosis of bALS.

2. Case series

2.1. Case 1

A 78 year old patient presented from an outside facility describing a progressive dysphagia to liquids and lingual dysarthria over one year. Neurological evaluation was remarkable for left hemi-tongue atrophy, fasciculations, and limited lingual mobility. Electromyography (EMG) of the right hemibody, performed at the referring facility, was interpreted as normal. Repeat focused EMG revealed reduced recruitment (1+), fasciculations (1+) and variability of the motor unit action potentials within the left genioglossus muscle consistent with an isolated LMN process of the left hypoglossal nerve. Magnetic resonance imaging (MRI) demonstrated a 28 mm × 38 mm enhancing left tongue base mass Fig. 1. Lymph node biopsy was consistent with metastatic squamous cell carcinoma [stage III, T4aN2cM0 with human papilloma virus (HPV) family 16 positive]. This patient was subsequently referred to

oncology for consideration of chemoradiation and surgical options.

2.2. Case 2

An 81 year old patient with no pertinent previous medical history presented to the outpatient Neurology clinic for evaluation of speech difficulties. She described a 1 year history of progressive slurring of her speech and food trapping on the right side of her mouth when chewing. Neurological evaluation was remarkable for right hemi-tongue atrophy with fasciculations. A MRI of the brain revealed a synovial cyst encroaching on the right hypoglossal canal with extension inferiorly to C1 Fig. 2. She was then referred to neurosurgery for consultation regarding cyst drainage.

3. Discussion

Clinicians are often faced with patients describing symptoms of dysarthria and note tongue fasciculations and/or tongue atrophy on examination. Commonly, the presence of these LMN findings strikes fear into the heart of the examiner as the mind reflexively entertains a diagnosis of ALS. However, it is important to remember that any process that disrupts the hypoglossal nucleus, the axon, or the lingual muscle itself may mimic a LMN process such as bALS. Careful evaluation of patients with suspected LMN lingual dysarthria is required, as bALS has a poor prognosis of typically less than 2 years and a significant reduction in quality of life [4].

Neuroanatomically, the motor pathway for lingual movement originates in the primary motor cortex and reaches the hypoglossal

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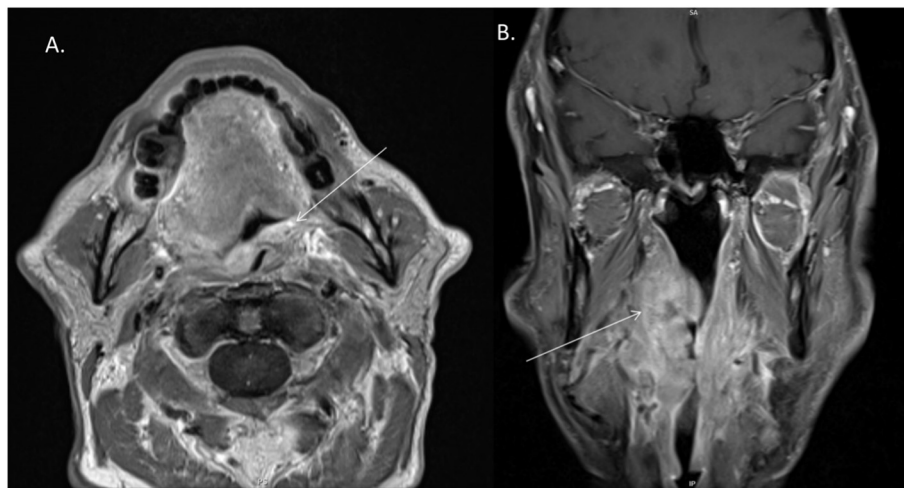


Fig. 1. Section A: Axial T1 post-contrast sequence. Section B: Coronal T1 post-contrast sequence. Magnetic resonance imaging revealing asymmetry of the tongue base with contrast enhancement with resultant pathologic diagnosis of squamous cell carcinoma.

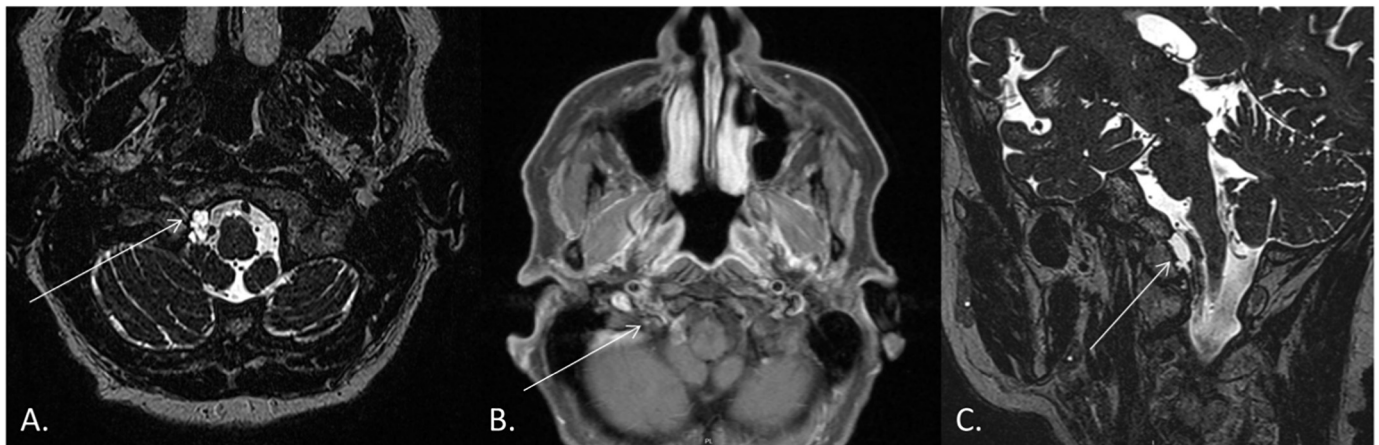


Fig. 2. Section A: Axial T2 SPACE sequence. Section B: Axial T1 post-contrast sequence. Section C: Sagittal oblique SPACE T2 sequence. Magnetic resonance imaging of the posterior fossa revealing a C1/occipital cyst with encroachment of the hypoglossal canal.

nucleus adjacent to the fourth ventricle in the medial medulla via the corticobulbar tracts. The hypoglossal nerve (cranial nerve XII) emerges from the ventral medulla inferior to the inferior olivary nuclei bilaterally and exits from skull base via the hypoglossal foramen (as seen in Fig. 2). The hypoglossal nerve then innervates all somatic intrinsic and extrinsic motor groups of the tongue, with the exception of the palatoglossus muscle. LMN lesions (CN XII nuclei and axon) create ipsilateral weakness with the tongue deviating towards the lesional side. Disruption of the hypoglossal nerve track, such as within lingual muscles (Case Report 1), or through encroachment of the hypoglossal foramina, (Case Report 2), will yield LMN pattern weakness, similar to that of bALS.

The differential diagnosis of LMN hypoglossal nerve palsy is extensive. The hypoglossal nucleus receives vascular supply through the perforator branches of the vertebral arteries and anterior spinal artery, making it vulnerable to infarction (medial medullary syndrome). However, this rarely presents with an isolated hypoglossal palsy as other vital structures are nearby. Within the hypoglossal canal itself, hypoglossal schwannomas, craniocervical junction juxta-articular cysts, non-articular/nonenhancing cysts, clival osteomyelitis, and Chiari type I malformation have been reported to cause LMN symptoms [5–7]. Further, compression of the hypoglossal nerve distal to the hypoglossal canal may occur rarely during intubation or in the presence of an infiltrative process (autoimmune versus malignancy) of the axon or intrinsic musculature of the tongue. Neurodegenerative processes may

present similarly; namely ALS, the spinocerebellar ataxias, and facial onset sensory and motor neuropathy (FOSMN syndrome). Rarely, systemic processes such as mitochondrial myopathy, anti-muscle-specific tyrosine kinase (MuSK) myasthenia gravis, and Lyme disease polyradiculopathy may present analogously [8–10].

In summary, patients presenting with LMN findings within the hypoglossal nerve distribution requires a broad differential for evaluation and diagnosis. Clinicians must not forget the potential implications that incorrect diagnoses may cause and so it is important for clinicians to remember: not all that fasciculates is ALS.

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