Isolated hypoglossal nerve palsy secondary to basilar artery compression: A case report and reviewing of electrodiagnostic evaluation of the hypoglossal nerve

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ABSTRACT

Introduction: Hypoglossal nerve palsy causes ipsilateral tongue weakness, commonly presenting with dysphagia, dysarthria, or perceived tongue weakness. Vascular compression is a rare cause of isolated hypoglossal nerve palsies. Imaging and serologic labs are common parts of the evaluation of hypoglossal nerve palsies. Though less commonly used, electrodiagnostic studies can be important in the diagnostic evaluation of hypoglossal nerve palsies.

Case: We report a case of a 53-year-old man with dysphagia found to have a left hypoglossal nerve palsy secondary to vascular compression from the basilar artery confirmed by electrodiagnostic and radiographic studies and we provide a review of the electrodiagnostic evaluation of the hypoglossal nerve.

Keywords: Vascular compression, isolated hypoglossal nerve palsy, basilar artery

Introduction

The hypoglossal nerve is a pure motor nerve innervating the intrinsic and extrinsic muscles of the tongue. It can be divided into 5 sections: medullary (nuclear), cisternal (extramedullary intracranial), skull base, nasopharyngeal/oropharyngeal carotid, and sublingual.[1] Hypoglossal nerve deficits that are distal to the nucleus cause ipsilateral tongue weakness. The most common initial symptoms of an isolated hypoglossal nerve palsy are dysphagia (38.5%), dysarthria (28.3%), tongue weakness (22.3%), headache (14.2%), and hoarseness (6.5%).[2] Isolated hypoglossal nerve palsies are rare. Causes of hypoglossal nerve palsies are extensive with common causes including carotid endarterectomy (15.2%), primary tumors (14.2%), including squamous cell carcinoma of the tongue, parotid tumors, tongue adenoid cystic carcinoma, and brain stem gliomas, metastatic tumors (13%) including lung cancer, renal cell carcinoma, rectal and pancreatic, prior radiation (6.2%), and inflammatory conditions (7.3%) including vasculitic multiple mononeuropathy, post-surgical inflammatory mononeuropathy, rheumatoid arthritis pannus, neurosarcoidosis, and Sjogren's syndrome. More rare causes of hypoglossal nerve palsies include trauma (4.1%), vascular (excluding post-op) causes (3.3%), congenital (2.8%), cystic (2.4%), and motor neuron disease (1.6%).[3]

Case presentation

A 53-year-old right-hand dominant male with a history of hypertension, hyperlipidemia, diabetes, and atrial fibrillation status post an atrial ablation 13 years prior, presented to the neurology clinic with a 4-5 month duration of difficulty initiating swallowing. He had previously been evaluated by an otolaryngologist and speech therapist with unremarkable modified barium swallow testing. His initial exam was significant for left tongue deviation both at rest and with protrusion, mild left-sided tongue atrophy without other cranial nerve deficits. His extremity muscle strength was normal, with decreased distal lower extremity reflexes and decreased sensation in a glove and stocking distribution. He had unremarkable serum inflammatory and autoimmune laboratory workup with normal erythrocyte sedimentation rate, C-reactive protein, antinuclear antibody, and rheumatoid factor as well as normal serum Lyme titers. A non-contrast head computed tomography (CT) scan was normal. Magnetic resonance imaging (MRI) of the brain with and without contrast (Figure 1) revealed mass effect of the basilar artery on the left upper medulla at the region of the left 12th cranial nerve without evidence of infarct or space-occupying lesions in the brainstem.

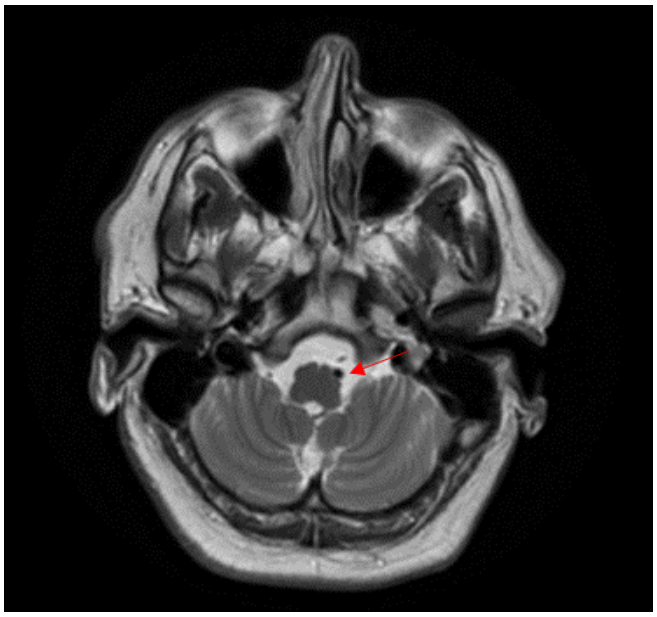
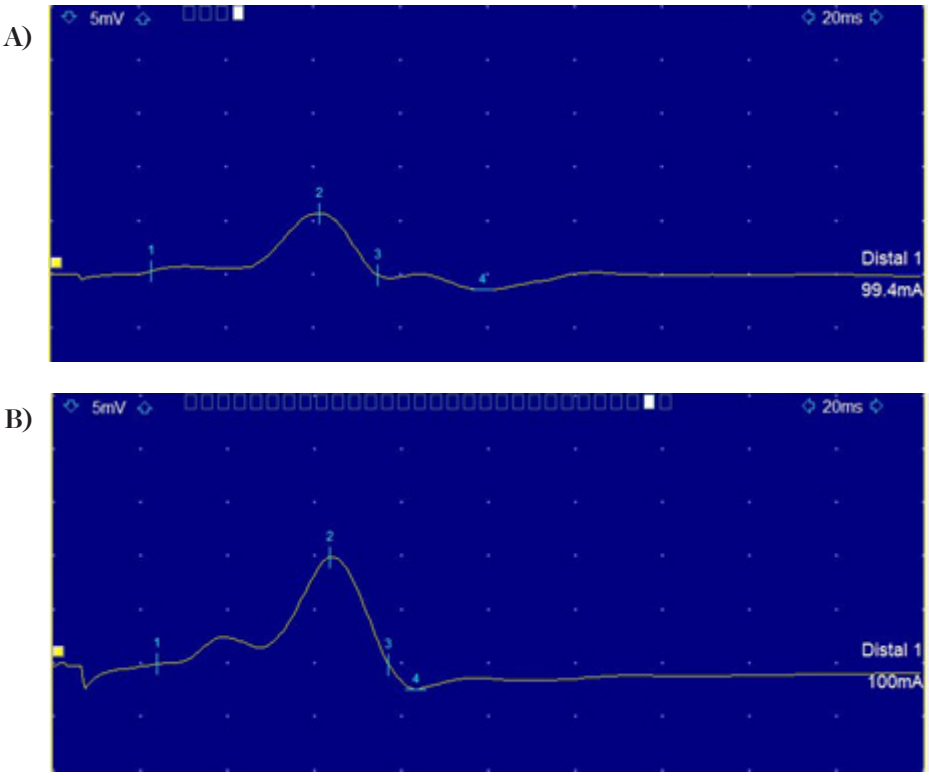


Figure 2: A) Left hypoglossal nerve conduction study at the intrinsic tongue muscle B) Right hyoglossus nerve conduction study at the intrinsic tongue muscle

On needle exam of the genioglossus muscle long duration and high amplitude motor unit action potentials (MUAP) were observed on the left without abnormal spontaneous activities .Needle examination of the right genioglossus muscle and left mentalis was normal. The facial motor NCS were normal and symmetrical bilaterally.



Subsequent nerve conduction studies (NCS) were completed by stimulating the hypoglossal nerve and recording over the dorsal surface of the tongue over the intrinsic tongue muscles. Findings were significant for relative decreased amplitude of the left hypoglossal motor nerve response compared to the right (right: amplitude of 10.0 mV with a latency of 2.4 ms, left: amplitude of 5.4 mV, latency of 2.3 ms) [Figure 2].

The patient had improvement in activation and protrusion of his left tongue between the initial exam and electrodiagnostic testing with persistent prominent tongue deviation. He continued to participate in speech therapy using compensatory and rehabilitative techniques and reported symptomatic improvement in dysphagia at a 6-month virtual follow-up.

Discussion

Vascular (excluding post op) causes of isolated hypoglossal nerve palsy are rare, accounting for 3.3% of all cases.[3] Vascular compression is a known non-surgical vascular cause of hypoglossal nerve palsy. Previous case reports detail hypoglossal nerve compression from persistent primitive hypoglossal artery[4] intracranial vertebral artery,[5] adherent fusiform PICA aneurysm[6] and basilar artery compression[7]. Treatment for vascular compression included spontaneous resolution or surgical decompression.

Initial evaluation of isolated hypoglossal nerve palsies entails evaluation of inflammatory, infectious, and compressive causes via serum and cerebrospinal fluid testing for inflammatory and infectious markers, and brainstem and cerebral vessel imaging. Though less commonly used, electrodiagnostic studies can be used to confirm and further evaluate hypoglossal nerve palsies. Electrodiagnostic evaluation of the hypoglossal nerve was first reported by Skorpil and Zverina in 1962 and later refined at Walter Reed Army Medical Center by Redmond and Di Benedetto in 1988.[8,9] Nerve stimulation is performed along the mandible, one-third from the angle of the jaw to the mental protuberance and 1 cm medial to the mandibles. The recording electrode is positioned 1 cm posterior to the lower incisors and the reference electrode is positioned 2

cm posterior to the recording electrode. The electrodes can be held on the anterior surface of the tongue via a special mouthpiece or a tongue depressor. Of the 30 subjects sampled, Redmond and Di Benedetto reported a mean latency of 2.2 +/- 0.4 ms (range 1.4-3.2 ms) and a mean amplitude of 3.8 +/- 1.6 mV (range 1.0-8.0 mV).[8] Needle electromyography of the genioglossus muscle can be used to evaluate hypoglossal nerve function, which is achieved by needle insertion intraorally or inferiorly via a subcutaneous approach from the medial angle of the mandible.[10] Electrodiagnostic testing can be easily completed on the hypoglossal nerve to assess motor function as an adjunct to serologic and radiographic evaluation of hypoglossal nerve palsies. This case reports an important cause of hypoglossal nerve palsies and reviews the potential utility in electrodiagnostic studies during evaluation.

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