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Electromyographic findings directing the diagnosis of an upper motor neuron lesion of the hypoglossal nerve

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Clinical image description

A 45-year-old woman presented with a subacute, intermittent deviation of the tongue to the right side, and dysarthria. Symptoms worsen and become more evident when the patient attempts to speak for a prolonged period of time. Neurological examination revealed normal cognitive function and memory. She had normal power, sensation, and reflexes in the four limbs. She had a normal gait and no limb or truncal ataxia. The examination of the cranial nerves was normal except for the deviation of the tongue to the right with mild atrophy. The dysarthria and tongue deviation were episodic, improving upon rest and worsening with talking. No dysphasia, diplopia, or dysphagia were reported. The clinical impression was that of a pathology of the right hypoglossal nerve or nucleus. The hypoglossal nerve is the 12th cranial nerve, exiting the brainstem at the level of the medulla and passing through the hypoglossal canal. It is purely a motor nerve innervating the ipsilateral intrinsic and extrinsic tongue muscles. A lesion of the hypoglossal nerve causes the tongue to deviate to the ipsilateral side because of the unopposed thrust of the healthy half of the muscle [1,2].

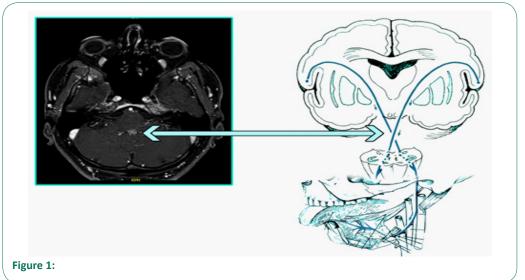
A supranuclear or supra-bulbar lesion above the hypoglossal nucleus results in the tongue deviating away from the lesion be-

cause of the neural crossing of the upper motor neurons. Supranuclear lesions do not cause atrophy contrary to infra-nuclear and nuclear lesions [1,2].

The concentric needle EMG examination performed on the patient's tongue revealed completely normal motor units and full recruitment on the left side. On the right side of the tongue, the needle would not pick up any spontaneous or voluntary motor units even when the patient tried to protrude the tongue maximally. These findings could not confirm a lesion of the hypoglossal nucleus or nerve, but rather a supra-bulbar lesion inhibiting activation of the contralateral part of the tongue.

High-resolution studies by MRI with gadolinium are always necessary for a successful imaging evaluation of hypoglossal disorders [3]. The patient underwent an MRI of the brain with contrast and thin cuts of the posterior fossa. This revealed an enhancing small lesion in the fourth ventricle at the area of the crossing of the supra bulbar innervation of the hypoglossal nerves. The lesion lies just posterior to the medulla. The suspicion of a malignant lesion prompted the necessary investigative studies revealing a primary breast cancer with bone and brain metastasis.

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