

Dysphonia of Central Origin

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DOI: [10.36347/sjmcr.2022.v10i02.030](https://doi.org/10.36347/sjmcr.2022.v10i02.030)

| Received: 09.01.2022 | Accepted: 13.02.2022 | Published: 28.02.2022

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Abstract

Case Report

Dysphonia is a frequent symptom in ENT outpatient clinic. In adults, apart from dysfunctional dysphonia, etiologies are linked to mucosal, muscular, joint or neural pathology of the vocal folds. The later can be of peripheral or central origin. We report the case of a patient with dysphonia in the context of amyotrophic lateral sclerosis pseudo-syndrom revealing an extra-axial ependymoma of the brainstem. A 35 years-old patient with no significant history presented to the outpatient clinic for dysphonia and chronic dysarthria recently complicated by dysphagia developing in a context of deterioration of general health condition. Physical examination found symptoms compatible with amyotrophic lateral sclerosis of bulbar onset syndrom. MRI identified an extra-axial tumor of the medulla-spine junction. Neurosurgeons performed a complete surgical removal through a suboccipital approach. Dysphonia due to brainstem involvement is rare. It should be considered before clinical signs of vagus nerve damage associated to another neural tract involvement.

Keywords: Amyotrophic lateral sclerosis, dysphonia, brainstem tumors, ependymoma.

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INTRODUCTION

Dysphonia is a usual ENT consultation motive that is often considered a trivial symptom. Mainly neglected at least at the beginning, patients seek help of the otolaryngologist when dysphonia is persistent or recurrent or associated to another sign such as dyspnea, dysphagia or cervical lymphadenopathy. In adults, beside dysfunctional dysphonia, dysphonia may be related to vocal folds pathology (laryngitis, benign or malignant tumor, myopathy....) or to a compromised innervation of peripheral or central origin. Yet uncommon, the central origin is often suspected before a single or multiple cranial nerve palsy associated to involvement of long neural tracts (motor or sensory) or cerebellar tracts. We report a case of central dysphonia associated to neurologic signs highly suggestive of amyotrophic lateral sclerosis which reveals an ependymoma of the junction between medulla oblongata and the spinal cord.

CASE PRESENTATION

A 35 years old male patient, with no personal history of smoking, drinking alcohol or drugs use and no family history of neurology disorder presented to the ENT outpatient clinic with a progressive onset of dysphonia and dysphagia that lasted for a year. The patient reported a recent occurrence of dysarthria and persistent fatigue with significant weight loss (10 Kg in 5 months). No limbs weakness, bladder or bowel dysfunctions or respiratory problems were reported. Also, no features of high intracranial pressure were found such as headache, vomiting or visual impairment. ENT examination disclosed left vocal fold palsy with fasciculations and amyotrophy of the left half of the tongue (Figure 1) with morphologically normal upper aero-digestive tract. Also, physical examination found symmetric pathologically vivid and poly-kinetic osteotendinous reflexes in upper limbs with amyotrophy prevailing at the left hand (Figures 2 & 3).



Figure 1: Amyotrophy and palsy of the left half of the tongue

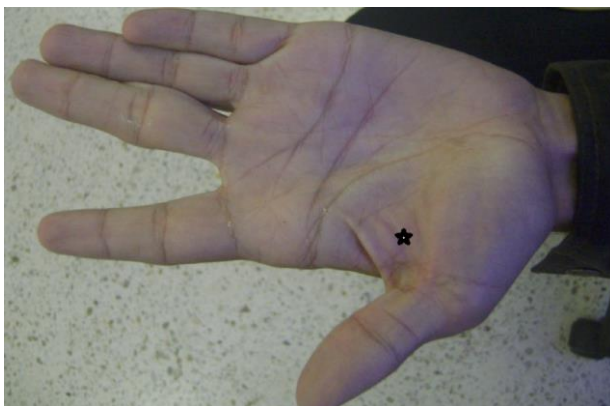


Figure 2: Amyotrophy of the left-hand muscles (star)



Figure 3: Amyotrophy of the left interosseous muscles

Otherwise, there were no gait disturbance, no spasticity, no other cranial nerve involvement and no sensory or cerebellar impairment. Clinical presentation was consistent with an involvement of upper and lower motor neurons that interest also cranial nerves referring to a bulbar onset of amyotrophic lateral sclerosis (ALS). Cerebral MRI performed as a routine test in ALS

assessment (Figures 4) has disclosed an extra axial brainstem tumor that is of an oblong shape involving the cranio-spinal junction from the left inferior cerebellar peduncle to the cervical spinal cord with significant swelling that infiltrates the medulla and the spinal cord.

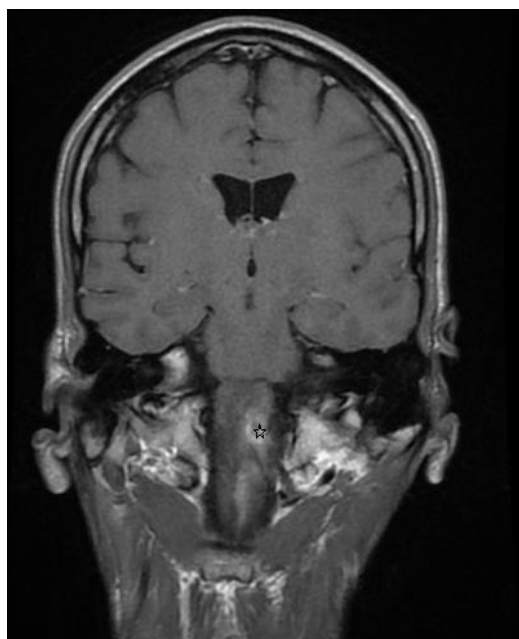


Figure 4: MRI Coronal T1 Gadolinium weighted image of the brainstem showing an heterogeneous enhancement of the tumor (star)

These findings were consistent with an ependymoma, or hemangioblastoma or a high-grade glioma. The patient underwent tumor resection through a suboccipital neurosurgical approach to the brainstem. Unfortunately, the patient died from complications of nosocomial pneumonia in the intensive care unit. The result of pathology examination of the surgical specimen was consistent with an ependymoma.

DISCUSSION

Dysphonia of neurological origin can be paralytic or dystonic. Paralytic dysphonia is due to any injury of the peripheral motor neuron all the way between the ambiguous nucleus and effector muscles. This might include the ambiguous nucleus, intramedullar neurons of vagus nerve, the vagus nerve main trunk or the recurrent nerve. One third of unilateral laryngeal palsy is idiopathic, 50% of these unilateral "idiopathic" laryngeal paralysis regress spontaneously within 6 months [1]. The other two main causes are related to surgery and / or cancer. The rest of neurological causes (less than 5% of cases) are responsible for unilateral but also bilateral vocal fold palsy such as Wallenberg syndrom, amyotrophic lateral sclerosis (ALS), myasthenia gravis, Guillain-Barre polyneuropathy, Charcot-Marie-Tooth disease, multiple sclerosis, central degenerative diseases, Lyme disease, inflammatory and infectious neuropathies (tuberculosis, HIV, sarcoidosis, diabetes, lupus, zona ...) [2].

ALS is a neurodegenerative disease which diagnosis is based on suggestive clinical findings of combined upper motor neuron and lower motor neuron involvement that cannot be explained by any other disease process, together with progression compatible with a neurodegenerative disorder [3, 4]. The mean age

of onset for sporadic ALS varies between 55–65 years with a median age of onset of 64 years [5, 6]. Only 5% of cases have an onset before the age of 30 years [6]. Bulbar onset is more common in women and in older age groups, with 43% of patients over the age of 70 years old presenting with bulbar symptoms compared to 15% below the age of 30 years old [6-8]. The onset and the early course of the disease are usually insidious. The diagnosis is sometimes suggested after 12 months or more of progression. Once the diagnosis is suspected, additional tests are rather made to rule out other curable conditions. Despite advances in investigative medicine over the past century, the diagnosis of ALS is based on the presence of very characteristic clinical findings in conjunction with investigations to exclude "ALS-mimic" syndromes. The World Federation of Neurology (WFN) Research Group on Motor Neuron Diseases have developed the 1994 'El Escorial' diagnostic criteria [9] and the revised 2000 'Airlie House' criteria [10] to aid in diagnosing and classifying patients for research studies and drug trials. According to the revised El Escorial Research Diagnostic Criteria for ALS [9] our patient had a "Probable ALS". Nevertheless, this case was slightly atypical as it presented an early start and slowly progressive sporadic bulbar onset ALS features.

As differential diagnosis, many skull base conditions and cervical myelopathies have to be taken into consideration such as Arnold-Chiari-type1 and other hindbrain malformations, foramen magnum lesions, intrinsic and extrinsic brainstem tumors or syringomyelia. However, in those cases other neurological symptoms like sensory loss or cerebellar syndrome may help sitting up the diagnosis. In our case, motor neurons involvement was the only complain. MR imaging disclosed a brainstem tumor and clearly

established the extra axial site of the tumor. Although no MR imaging features are pathognomonic of ependymoma [11], they often (in half the reported cases) show heterogeneous enhancement [11], as did this mass. This lack of imaging specificity most likely reflects the varied histology of these tumors [2]. Two to six percent of all primary brain tumors are ependymomas [12, 13]. These neoplasms are gliomas derived from ependymocytes that tend to present as malignant intracranial tumors in children and as benign intraspinal neoplasms in adults. It may arise from any level of the ventricular system or central canal of the spinal cord. Intracranial ependymomas most frequently originate in the fourth ventricle and histologically are densely cellular tumors which may contain ependymal tubules and perivascular pseudo-rosettes [14, 15]. An exclusively extra axial ependymoma of the posterior fossa is rare. We are aware of two other reported cases [16, 17].

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