Progressive bilateral tongue atrophy: a case of misdiagnosed motor neuron disease

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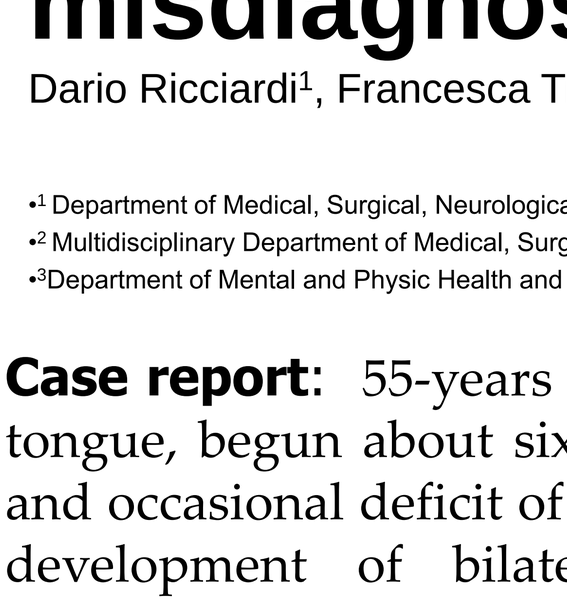
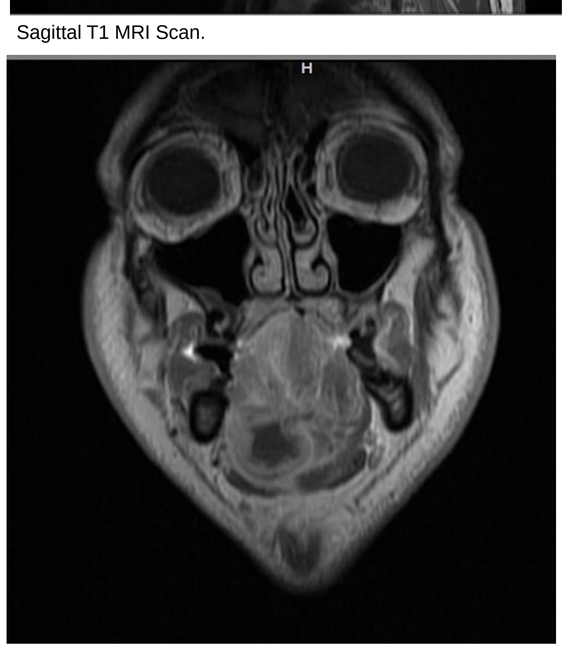
Case report: 55-years man who, firstly, exhibited a left hemiatrophy of the tongue, begun about six-years before and characterized by speech difficulty and occasional deficit of swallowing, with progressive motor impairment and development of bilateral atrophy. It was hypothesized an atypical presentation of motor neuron disease with prominent involvement of the bulbar site. In favour of this hypothesis there was the detection of slight T2 and FLAIR hyperintensities of the cortico-spinal tracts at magnetic resonance imaging (MRI) of the brain. However, we evaluated all the potential differential diagnoses (1-3), including traumatic brain injury, tumours, infections and autoimmune, endocrine or vascular anomalies.

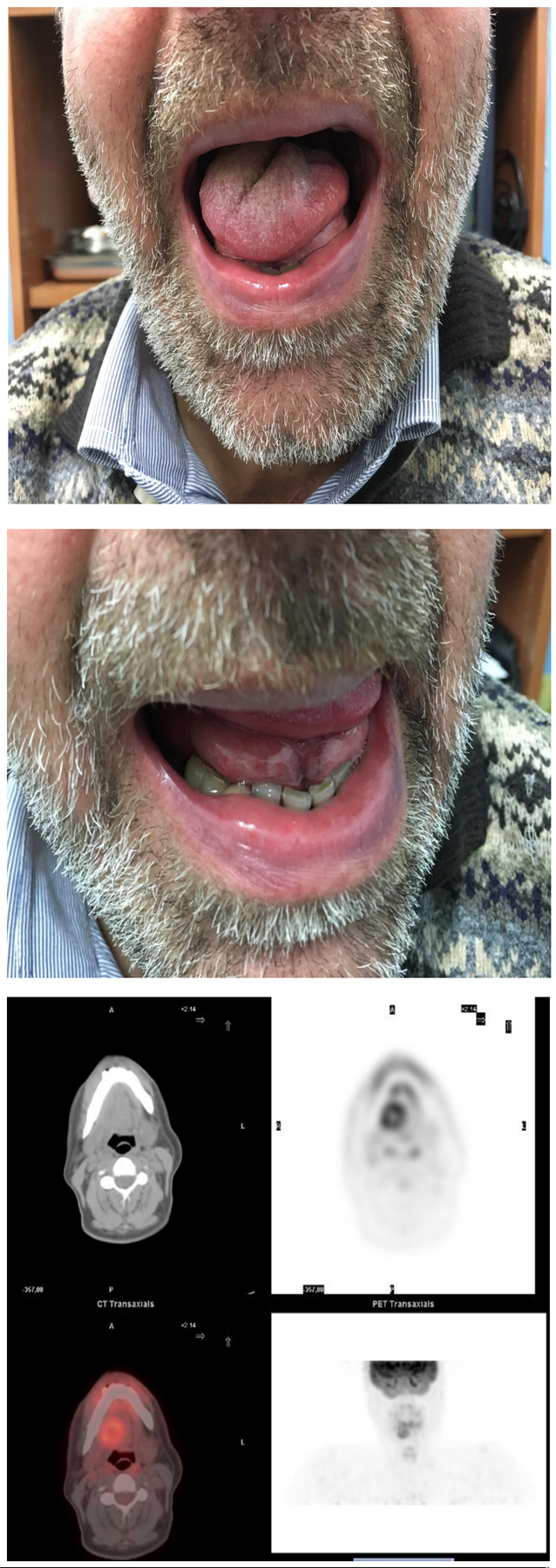


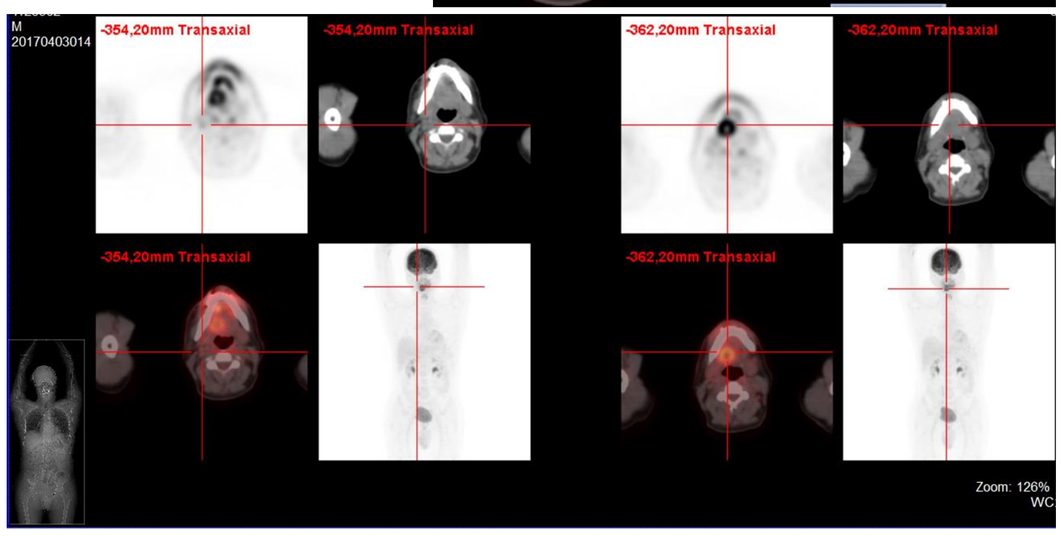
Sagittal T1 MRI Scan.



Coronal T1 MRI Scan.

Results: Among instrumental findings, the electromyography and motor evoked potentials, performed at bulbar and limb sites, did not show any abnormality related to upper or lower motor neurons impairment. Conversely, head and neck MRI, performed before and after administration of gadolinium, revealed a keratin cyst under the tongue and symmetrical fatty striations, atrophy, and fibrosis of all the tongue with right-side enhancement after gadolinium, potentially consistent with a neoplastic lesion. Finally, the oral mucosa and tongue biopsy showed cellular abnormalities consistent with CKHW+, CK7+ and p63+ squamous cell carcinomas of the tongue. Whole-body 18f-fluorodeoxyglucose positron emission tomography revealed a slight uptake of the tracer at the right lateral-cervical lymph nodes.





Whole body Pet-18 FDG

Conclusions: Our patient underwent an oncologic assessment and was addressed to begin chemotherapy and radiotherapy. The neoplastic hypothesis should be always considered in cases of atypical presentation and progression of bulbar signs. Moreover, MRI seems to be able to visualize corticospinal tract abnormalities, even if not sensitive for motor neuron diseases.

•**Bibliography**
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