CLINICAL AND RADIOLOGICAL FOLLOW UP IN HYPOGLOSSAL NERVE MONOPARESIS IN COURSE OF SJÖGREN'S SYNDROME (SS) AND SCLERODERMIA (SSC)

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Introduction: *Hypoglossal nerve lesions may result in subsequent ipsilateral paralysis and atrophy. Various ethiologies can be advocated (traumatic, neoplastic, vascular, inflammatory). Traction, direct compression, tissue invasion or vascular damage can all determinate the deficit. In some cases multiple low cranial nerve (IX, X, XI, XII and sympathetic nerve) involvement configures various clinical pictures such as Collet-Sicard, Villaret or Jackson syndrome. Isolated cranial nerve involvement in Sjögren's syndrome (SS) or sclerodermia (SSc) has rarely been described but no report on isolated hypoglossal nerve monoparesis has been described in co-existing connective tissue disorders.*

Case Report: *62 y-o woman (Fig1), presenting with isolated hypoglossal nerve paresis. MRI (Fig2) and CT scan showed only periodontal pannus without calcifications, contrast enhancement or signs of vascular or brain stem involvement by the mass or atlanto-axial dislocation. EMG was normal. Few mounths later she devellopped hypoglossal neuvralgic pain, well controlled by pregabalin 150 mg/die. She did not have history of peripheral arthritis or arthralgia, but reported sicca symptoms and recent onset Raynaud's phenomenon. Laboratory tests disclosed rheumatoid factor (low titler), anti-SSA, and anti-centromere antibodies positivity, while anti-citrullinated peptides were absent. Schirmer's test was pathological and scleroderma pattern at nailfold videocapillaroscopy examination. She satisfied the American College of Rheumatology criteria for both SSc and SS. She didn't start any therapy for the reumathologic deasese because of her poor systemic symptomatology. However, after 12 months the rheumatological and neurological symptoms spontaneously regressed and also periodontal pannus at the MRI was reduced (Fig3).*

Discussion: *The presence of pannus is a typical characteristic of long-standing rheumatoid arthritis but, for our knowledge, it is the first description in Sjögren's syndrome and sclerodermia. Moreover, our patient presented with rare systemic symptoms and her clinical history started with a severe radiological injury but an isolated neurological symptom, that regressed without therapy. In conclusion, neurological complications of systemic disease must be consider also in case of paucisymptomatic presentations and a rheumatological evaluation is mandatory for a correct diagnosis. The pannus in Sjogren's syndrome and sclerodermia may have a different pathogenetic mechanism than in rheumatoid arthritis and we need more description and follow-up for a best knowledge of its characteristics and evolution*

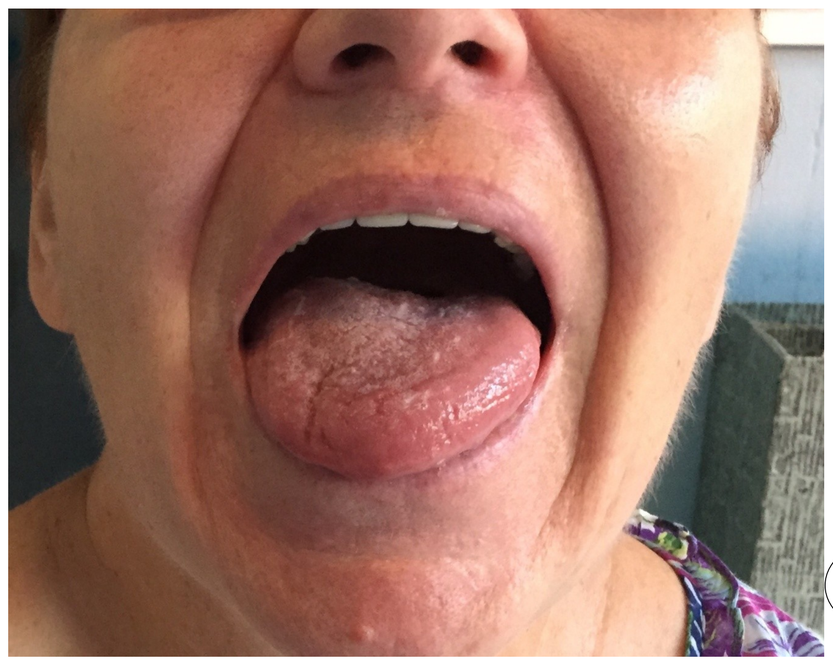

Fig 1

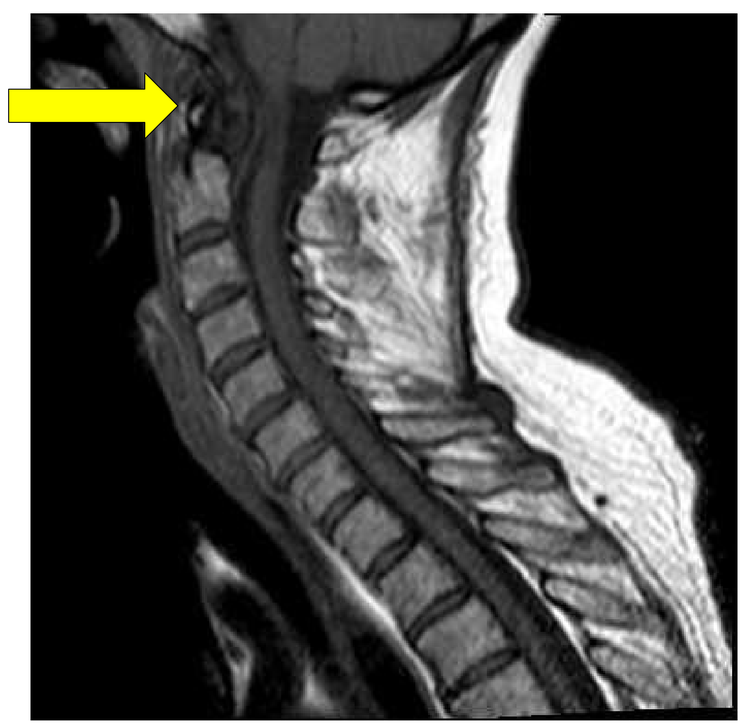

Fig 2

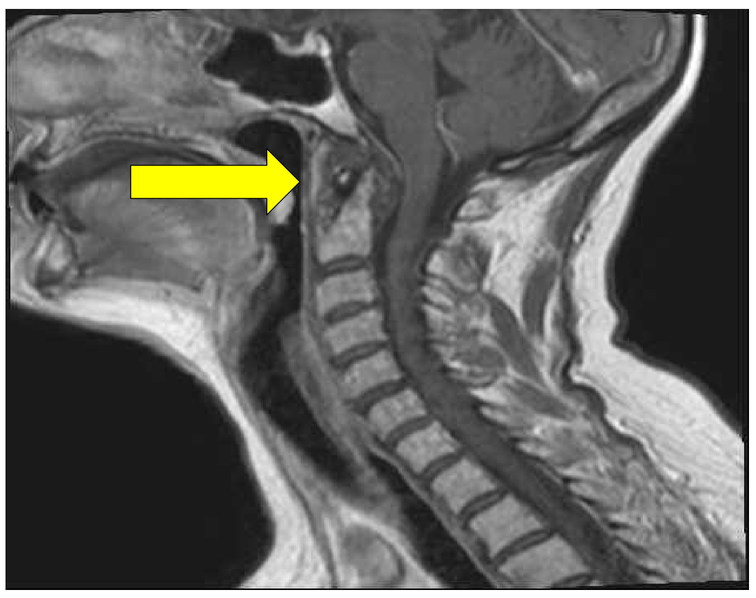

Fig 3

Conclusions: *The cervical mass did not show the typical features of rheumatoid pannus and not even of crowned dens syndrome and seems to be reduced over time. Its clinical evolution correlates with the radiological amelioration. Therefore, our previous clinical hypothesis that isolated hypoglossal nerve monoparesis occurs during a connective tissue disease, due to the traction, dislocation and compression of the periodontal pannus, seems to be correct.*

References:

Lee P, Bruni J, Sukenik S Neurological manifestations in systemic sclerosis (scleroderma). *The Journal of Rheumatology (1984);11:480-3*

Teasdall RD, Frayha RA, Shulman LE Cranial nerve involvement in systemic sclerosis (scleroderma): a report of 10 cases. *Medicine (1980);59:149-59*

Ashraf VV, Bhasi R, Kumar RP, Girija AS Primary Sjögren's syndrome manifesting as multiple cranial neuropathies: MRI findings. *Annals of Indian Academy of*

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