MELKERSSON-ROSENTHAL SYNDROME: A CASE REPORT

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Case Report

We describe the case of a 39-years-old woman presenting to our attention for a newly onset left-sided, lower motorneuron type, facial nerve palsy. Neurological examination didn't revealed any significant finding; a peculiar fissured tongue was observed (see Figure 1). Patient past medical history was remarkable for three previous episodes involving the contralateral 7th cranial nerve. A brain MRI performed at the time of the first episode revealed a tiny lesion gad-enhancing located along the facial nerve course. Each episodes were treated with oral steroids and resulted in full recovery. No family history of similar complaints was reported. An autoimmune thyroiditis was diagnosed on the basis of blood tests (thyroid-stimulating hormone 6,02 mcUI/ml, thyroperoxidase antibody >2000 IU/ml, thyroglobulin antibody 55,86 IU/ml) and thyroid ultrasound exam (demonstrating a coarse, heterogeneous, hypoechoic parenchymal echo-pattern). Serum blood tests for other autoimmune disorders and infectious diseases were normal. So it was cerebrospinal fluid examination. Electrophysiological studies confirmed a stabilized neuropathic damage involving facial nerves of both sides. Brain MRI was unremarkable. To rule out sarcoidosis and other interstitial lung disease a chest CT scan was performed and returned normal. A Melkersson-Rosenthal syndrome (MRS) was hypothesized as responsible of the recurrent facial nerve palsies. Patient was managed with prednisone 75 mg daily tapered over a 4-weeks period. Treatment led to partial recovery.

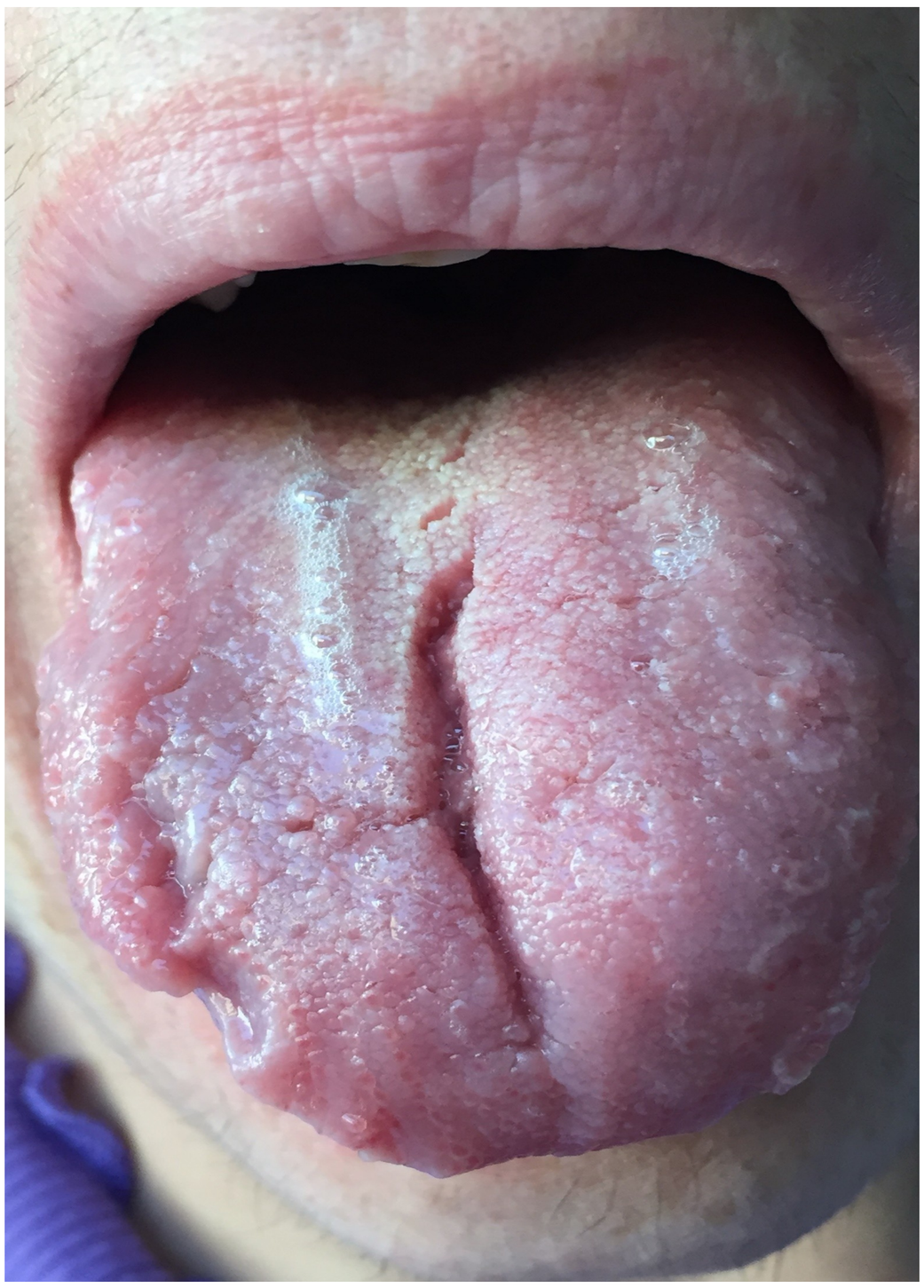

Fig.1 – Our Patient fissured tongue (*lingua plicata*)

Discussion

MRS is a rare cause of recurrent peripheral facial nerve palsy, typically characterized by the triad of orofacial edema, a peculiar fissured tongue (*lingua plicata*) and recurrent lower motorneuron facial paralysis. It has to be mentioned that the complete triad is rarer in presentation than oligosymptomatic forms. In a retrospective study of biopsied cases (1) the full triad was observed in 9 out of 72 patients (13%). Etiology is unclear; infectious, inflammatory, immunological and genetic causes have been advocated. Co-occurrence of MRS and autoimmune disorders has been documented and possibly argue in favour of an immunological pathophysiology. MRS diagnosis is generally applied to cases for which no other etiologic explanation can be identified; as in Our Patient, MRS and autoimmune thyroiditis co-existence have been rarely reported in literature (2). Some Authors propose that a definite diagnosis requires histological demonstration of non-caseating granulomas on skin biopsy. Treatment with anti-inflammatory drugs, oral steroids, antiviral drugs and immunosuppressive agents have been proposed with no consistent treatment responses (1). Prophylactic nerve decompression in selected patients may prevent future facial palsy and high-resolution CT of temporal bone may help in sorting out the most suitable candidates (3).

Bibliografia

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