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Case of Rare Entity - Parry-Romberg Syndrome

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3. Introduction

Parry-Romberg syndrome (PRS), also known as progressive facial hemyatrophy is a rare clinical entity with prevalence of 1 case on 250 000 of general population [1-3]. It was first described by C.Parry in 1825 and – later – by M.Romberg in 1846 [1,2]. Course of this syndrome ends in atrophy of subcutaneous fat with skin changes, *en coupe de sabre* sign, in some cases facial muscles wasting, ophthalmic and nervous systems involvement [4-9]. Exact cause & mechanism of these changes stays unknown; disruption of immunologic reactions seems to play significant role here [10,13]. SPR are more often seen in females, usually starts at the first decade of life and went on its clinical course for 2-20 years; it's a self-limiting condition [8].

There is a certain disagreement on a typical clinical, neurophysiologic and MRI features of the syndrome. Neurologic symptoms are described in 15-45% cases of SPR; ophthalmic involvement – in about 35% of the cases; sometimes *en coupe de sabre* may be located on the neck rather than face, or even absent; degree of the facial changes also vary [11, 15, 16]. MRI reveals changes in the brain in about 75% of the cases, although they are usually unspecific [5]. As about neurophysiology, spectrum of the findings is wide and different in different cases. Thus, some authors describe conduction disruption along *n.Facialis*, but some don't see it; same thing goes about blink reflex changes; periorbital muscles involvement according to the needle EMG; pain-evoked and brainstem auditory evoked potentials [19,21-25]. So, every description of this syndrome is somewhat valuable for its future research.

We present the case of a PRS in a female, Caucasian race, aged 43. First signs developed when she was 20 years old, without any specific health problems in previous life; symptoms started from the darkening of the skin on the forehead with a slow progression in the next 10 years; classic *en coup de sabre* sign appeared, with total wasting of subcutaneous fat tissue on the left side of the face (**Figure 1**).

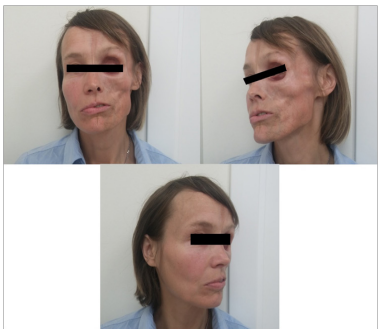

Figure 1. Facial changes in 43-years old female patient with Parry-Romberg syndrome.

Our patient had total blindness on the left eye, according to the ophthalmologist examination. According to the conduction studies (Neuro-MVP system, Neurosoft, Russia), including here the vegetative nervous system examination, there were no deviations from the normal data (**Figure 2** & **3**).

Needle myography of the m.Orbicularis Oculi et m.Orbicularis oris also obtained normal data.

Infrared thermography revealed changes of the facial temperature on the side of the lesion with the local hypothermia over the left cheekbone (**Figure 4**).

Brain MRI (1.5 Tesla *Philips Ingénue* device) revealed left enophthalmos, subcutaneous fat wasting, hypotrophy of the left *m. Masseter*, left-eyed wasting of the intraorbital fat and

unspecific periventricular lesions in the brain (**Figure 5**).

Head CT (Philips 128 *Ingenuity* device) with a 3-D reconstruction revealed no bones involvement (**Figure 6**).

Attempts to treat the condition with lipofilling procedures were undertaken twice and failed both times; all additional fat tissue degenerated in 2 and 3 months, accordingly. Attempts to treat the deformities would be taken with a reconstructive surgery (subcutaneous silicone fillings) and left eye prosthetics applications.

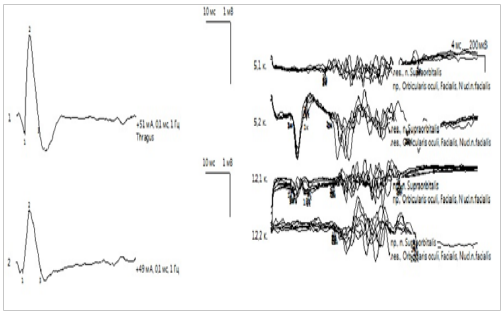

Figure 2. Facial nerve conduction & blink reflex data in a 43-years old female patient with Parry-Romberg syndrome.

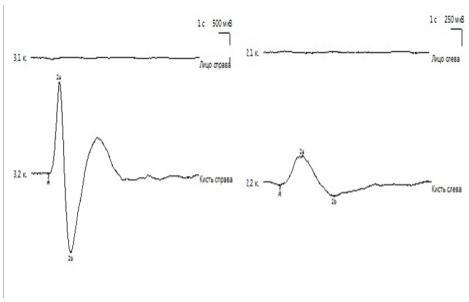

Figure 3. Galvanic skin test data in a 43-years old female patient with Parry-Romberg syndrome.

Infrared thermography revealed changes of the facial temperature on the side of the lesion with the local hypothermia over the left cheekbone (Fig. 4).

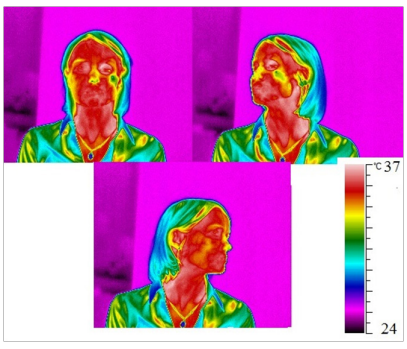

Figure 4. Infrared thermography of the 43-years old female patient with Parry-Romberg syndrome.

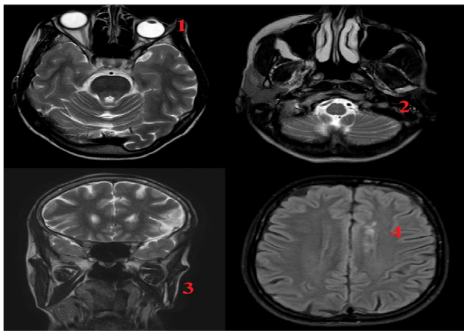

Figure 5. Brain MRI of the 43-years old female patient with Parry-Romberg syndrome. 1: enophthalmos, left-eyed wasting of the intraorbital fat; 2 – left parotid gland atrophy; 3 – hypotrophy of the left m.Masseter and subcutaneous fat tissue; 3 – unspecific periventricular lesions in the brain.

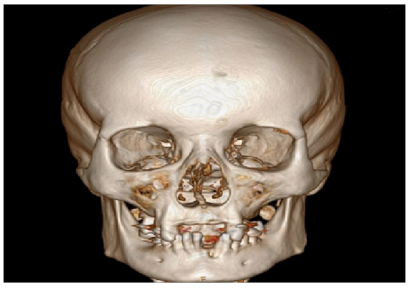

Figure 6. Brain CT of the 43-years old female patient with Parry-Romberg syndrome.

Thus, here we present a rare clinical entity of Parry-Romberg Syndrome with monolateral far developed and final deformations of the face and eye involvement. Contrary to some reports [23-25] conduction studies, galvanic reflex and needle EMG findings in our case were all normal. Thermography changes also may not be considered as a severe ones; as about MRI findings, we have found some brain lesions, and typically they were, according to the current knowledge, unspecific [5].

This wide spectrum of the symptoms and findings in different reports of PRS may be explained, probably, by its different etiology. Some cases of PRS are secondary to vascular processes, tumors *etc.*; others may represent genuine disease of yet unknown nature.

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