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PARRY-ROMBERG SYNDROME - CASE REPORT

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CASE PRESENTATION

A 25-year-old female referred to the vascular neurology outpatient clinic after hemorrhagic stroke in July 2021. She complained of daily pulsatile headache associated with nausea and left hemicranial location. Besides progressive deformity in the right hemiface.

Physical examination: good general condition, conscious and oriented, afebrile, acyanotic, anicteric and eupneic. Besides left hemiparesis and deformities in the skull, this one being hypotrophy in the right hemiface. Being requested Magnetic Resonance Imaging of skull and subsequently having the confirmation of the diagnosis of Parry-Romberg Syndrome.

DISCUSSION

The Parry-Romberg Syndrome (PPR) or progressive facial hemiatrophy is a disease of unknown origin, usually develops in the first or second decade of life, prevailing female gender, most often affects only one hemiface, as in the patient's report. It is manifested by a craniofacial atrophy that can affect part or all facial structures from skin, muscle, fat, bone tissue, trigeminal nerve and even the brain parenchyma (PRADO et al., 2011).

The SPR can affect the central nervous system, among the neurological manifestations, headache and seizure disorders are the most common. Neuropathies involving several cranial nerves (the third, fifth, sixth and seventh), vascular inflammation and destruction of the bone surrounding the trigeminal nerve causes secondary trigeminal neuralgia in these patients, manifesting with chronic facial pain that is usually resistant to treatment (ARIF et al., 2020)

Computed tomography and MRI are methods for diagnosing CNS changes, with the characteristic findings being: white matter hypersignal on T2 and FLAIR, leptomeningeal enhancement, and brain atrophy, being found on the patient's skull MRI. The differential diagnosis includes diseases that course with cerebral hemiatrophy, such as Rasmussen's encephalitis and Sturge-Weber syndrome, but in these we did not observe the typical hemifacial changes found in Parry-Romberg syndrome (PAULA et al., 2014).

FINAL COMMENTS

The reported case and exposed publications bring a rather rare diagnosis. As the patient's facial involvement has not yet significantly affected the quality of life of the patient, the professionals responsible for the case did not prescribe a surgical approach.