

# Parry-romberg syndrome: potential benefits of cannabinoid therapy - case report

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## Background

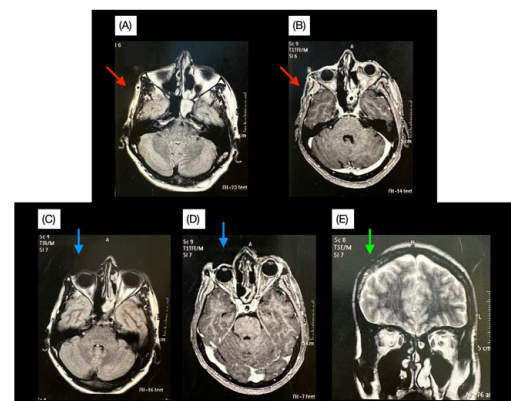
Localized scleroderma (SsL) is a connective tissue disease of unknown etiology, in which excess collagen deposition leads to the development of fibrotic lesions. Its clinical spectrum is heterogeneous, with multiple phenotypes (plaque, linear, among others). The coup de saber lesion (CS) is characterized by a unilateral sclerotic band that involves the scalp and the frontoparietal region, mostly without extension below the eyebrow implantation line. Parry-Romberg Syndrome (PRS) is considered a variant of CS, being a rare phenotype, characterized by progressive hemifacial atrophy, affecting skin, muscles, bones and structures of the central nervous system (CNS). Manifestations may include headache, trigeminal neuralgia and seizures. This study aims to report the therapeutic potential of the use of cannabinoid therapy in neurological manifestations in PRS.<sup>1-3</sup>

## Case report

Male patient, 40 years old, diagnosed at 31 years of age with PRS, presenting atrophy in the entire right hemiface, extending to the right parietal region and lagophthalmos. He used penicillamine 1 g/day for 6 months and methotrexate 20 mg/week for 5 years, in addition to aesthetic procedures, a few years after diagnosis. Undergoing ophthalmological follow-up using artificial tears. He began follow-up with a complaint of recurrent and disabling holocranial headache.<sup>4</sup> Brain MRI showed enophthalmos on the right, atrophy of the subcutaneous tissue and of the masseteric and temporal muscles on the right, in addition to foci of signal alteration in the periventricular white matter of the right cerebral hemisphere. The headache was chronic, disabling and refractory to the continuous use, for more than 1 year, of amitriptyline 75 mg/day and valproic acid 2 g/day, either alone or in combination. The patient had a Headache-Related Disability Index (HDI-Brazil) score of 88 and a Headache Impact Test (HIT-6) score of 72, generating a significant negative impact on quality of life. *Cannabis sativa* promedol extract 200 mg/mL (CBD\* + THC\*\* combination) was prescribed at a dosage of 35 mg of CBD per day (7 drops every 8 hours), in combination with valproic acid 2 g/day and amitriptyline.<sup>5</sup> After 3 months of continuous use of *Cannabis sativa* promedol extract associated with the anticonvulsant, both questionnaires HDI-Brazil and HIT-6 decreased to 56 points, with a significant improvement in the patient's quality of life. After 7 months of using the *Cannabis* extract, there was a decrease in the dose of valproic acid to 1.5 g/day, without compromising the improvement acquired (Figure 1-3).<sup>6,7</sup>



**Figure 1 & 2** Patient with loss of symmetry of the right hemiface.



**Figure 3** (A) and (C) T2-Flair MRI showing myocutaneous atrophy, involving masseter and temporal muscles (red arrow) and enophthalmos (blue arrow). (B) and (D) T1 MRI with contrast showing facial asymmetry (red arrow) and enophthalmos (blue arrow). (E) Represents coronal-T2 plane, showing cutaneous/subcutaneous atrophy (green arrow).

## Conclusion

This case report demonstrates the therapeutic potential of cannabinoids in the neurological symptoms of a potentially disabling chronic disease, where therapeutic options are limited and little known.

## Acknowledgements

None.

## Conflicts of interest

The authors declare that there are no conflicts of interest.

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