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THE RARE AND IMPACTING PARRY-ROMBERG SYNDROME: A CASE REPORT.

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BACKGROUND

Parry-Romberg syndrome (PRS), a disease that has been described for more than a decade, also known as progressive hemifacial atrophy, is a sporadic neurocutaneous disease, characterized by a slow and progressive atrophy of the hemiface tissues, which can affect all tissues and also manifest neurological and ocular changes. This report is a 32-year-old patient who claims to have right hemifacial atrophy since childhood and findings suggestive of PRS in nuclear magnetic resonance (MRI) of the skull.

CASE REPORT

Female, started atrophy in right hemiface at 08 years of age, progressing with worsening of the condition over 24 years and, for aesthetic purposes, was referred from the basic health unit to the neurologist. Investigated with MRI that demonstrated cortical thickening and foci of hypersignal in T2 and FLAIR in the frontoparietal white matter on the right, findings considered more sugestive of the syndrome. Requested the screening of infectious diseases, tuberculin skin test, chest X-ray for the initiation of immunosuppression and referred to the dermatologist for cutaneous biopsy that showed superior dermis with dilated vessels, scarce perivascular lymphoplasmacytic infiltrate, sweat glomeruli with scarcely suppressed adipose cushion, remaining of the reticular dermis thickened and hyalinized bands of collagen, involving nervous fillet and invading hypodermis. Description suggestive of scleroderma. With the specific findings, laboratory tests without alterations, started methotrexate and referred to plastic surgery for adipose grafting in the right hemiface.

CONCLUSION

PRS is a rare pathology of unknown etiology, diagnosis is suggested by clinical history and imaging studies with more specific findings. Cutaneous biopsy may reveal scleroderma since there is sclerosis present in the middle and lower thirds of the face. An important differential diagnosis is linear scleroderma "en coup de sabre" deformity limited to the middle and lower thirds of the face and absence of alopecia. Surgical treatment is graded on a severity scale and only the fat graft of hemiface is proposed by plastic surgery.