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A Tale of Two Diseases: Parry Romberg Syndrome and En Coup De Sabre: A Case Report

Muhammad Erfan, Ikram ullah khan, Syed Afaq Ahmed, Saqib Hasnain
Department of Dermatology Pakistan Institute of Medical Sciences Islamabad

Abstract

Background: Parry Romberg syndrome is a very rare disorder of unknown etiology. A degenerative acquired disorder characterized by hemifacial atrophy. It can present with skeletal, dental and soft tissue abnormalities with or without CNS symptoms. Majority of the affected patients have manifestations of the disease in the first
two decades of life. Many cases have been documented having combination of Parry Romberg syndrome and another form of localized scleroderma- en coup de sabre morphea. Histological features of the disease include fibrosis with atrophy of adipose tissue and lack of any inflammatory infiltrate. Limited treatment options make the disease difficult to manage. Case presentation: We present a case of an otherwise perfectly healthy, 12 years old girl with early features of hemi-facial atrophy and en coup de sabre morphea.

Conclusion: Both Parry Romberg syndrome and en coup de sabre morphea have many localized and systemic associations. In many case reports neurological and dental abnormalities have been described. In our case patient did not appear to have any of these.

Keywords: Parry Romberg syndrome, hemifacial atrophy, en coup de sabre morphea.