Parry-Romberg syndrome and En Coup de Sabre overlap

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Abstract: Parry-Romberg syndrome (PRS) and En Coup de Sabre (ECDS) type of morphea has been reported sporadically in literature. We report a 30 year old female patient who presented with a purplish pigmented sclerotic plaque on the right side of face and scalp with obvious facial asymmetry. Additionally she had facial muscle tightness, migraine and myopia in the right eye. A provisional diagnosis of Parry-Romberg syndrome with En Coup de Sabre of morphea was made.

Keyword: "Parry-Romberg syndrome", "Progressive facial hemiatrophy", "En Coup de Sabre".

Introduction: Parry-Romberg syndrome (PRS) or Progressive facial hemiatrophy is a condition characterized by unilateral atrophy of subcutaneous tissue, muscle and occasionally bones, usually along trigeminal nerve distribution. It can affect brain, skull, hair, skin, eye, ear, nose, mouth, teeth, tongue and neck. Rarely it can affect the trunk as well. Linear scleroderma of frontal or fronto-parietal region is known as "En Coup de Sabre (ECDS)" a French phrase that described the injuries of foot soldiers struck on the head by the sword of a horse riding cavalry soldier. ECDS is characterized by purplish brown streak or a single white, atrophic band running vertically across the forehead. There is only a fine line of difference between these two conditions. In PRS skin is less bound down than that of in ECDS. Though sclerosis is not a presenting feature in any stage of PRS, morpheaform sclerosis was found to be present in 25% to 75% cases of PRS. Involvement of tongue and deeper atrophy usually favours PRS. Typical lesion of ECDS does not extend beyond the eyebrow, affects only upto forehead whereas PRS can affect the entire side of the face. PRS usually become inactive in 3-5 years leaving behind minimal atrophy with hyperpigmentation. Case report

A 30 years old female presented to our outpatient department with hyperpigmented skin lesions over right side of the face for the past 6 years. She complained of disfigurement of face along with blurring of vision in the right eye, severe headache and tightening of jaw muscles on the right side which worsened during emotional stress. There was no history suggestive of seizures or family history of similar lesions. On examination she had a linear sclerotic hyperpigmented plaque on the right side of the face extending from the fronto-parietal area to the chin. The affected area was difficult to pinch.

Biopsy of lesion of the forehead region showed thinned out epidermis. The dermis showed homogenous dense collagen deposition extending from papillary dermis and down up to the subcutaneous fat. The hair follicles and sweat glands were reduced. ANA was positive in 1:10 dilutions. Rheumatoid factor was positive in 16 U/ml dilutions. Ophthalmic examination revealed 6/24 vision in right eye and 6/6 in left eye. CT facial bones study was normal except for atrophic subcutaneous tissue. All other routine investigations were normal including CT brain.
Discussion Both Parry-Romberg syndrome and En Coup de Sabre are rare conditions of unknown etiology, with variable neurological involvement. The two processes may co-exist and it is mostly confused in literature. PRS usually affects deeper structures, while ECDS mainly affects superficial structures. In this case, the patient had increased pigmentation, right sided facial muscle spasm, myopia and atrophy along with atrophic facial subcutaneous tissue in CT which favours PRS. Purplish pigmented sclerotic plaque involving scalp, forehead, cheek and chin with histopathology showing homogenous hyalinised dense collagen throughout the dermis favours ECDS. In a classical case of PRS the skin may be normal and mobile. Here there is difficulty in pinching the skin. Thus we concluded this case as an overlap of ECDS and PRS. There are studies showing that typical linear scleroderma cases especially En Coup de Sabre turning into Parry-Romberg Syndrome after several years\textsuperscript{11}. In a case series of Progressive facial hemiatrophy by Feroze et al\textsuperscript{2}, none of them had purplish hue or histopathological evidence of sclerosis. Auvinet et al\textsuperscript{9} showed severe enophthalmos with eyelid atrophy, loss of cilia and mild blepharoptosis in a similar case of overlap. Here in our case, loss of eyebrows and eyelashes in the medial side of right eye was present and the patient had myopia in the right eye also. The various proposed etiologies for PRS include trauma, infections (Borrelia burgdorferi)\textsuperscript{4}, autoimmune mechanisms, genetic factors, etc. Parry-Romberg syndrome is more common in women\textsuperscript{3}. Though earlier age of onset is said to be common, later onset around 2-3\textsuperscript{rd} decade has also been reported. From various studies it has been shown that ECDS and PRS overlap is seen in 36-42\% of patients. Bony and neurological abnormalities have been associated with both the conditions. We can consider both ECDS and PRS as different presentations of same spectrum of disease. Various treatment options include topical Calcipotriol & low dose UVA1 phototherapy\textsuperscript{4}, topical Halofuginone\textsuperscript{10} and systemic drugs such as Methotrexate, Cyclophosphamide, Cyclosporine, D-penicillamine, Hydroxychloroquine\textsuperscript{3}. But none is effective consistently\textsuperscript{8}. Reconstructive surgeries using autologous fat transfer can be considered in disfiguring lesions.

Conclusion:
This case is presented here to highlight the presence of overlap of ECDS and PRS. Due to the close similarity between PRS and ECDS and the presence of atrophy in the late stages of ECDS and also the occurrence of morpheaform sclerosis in late stages of PRS, it is difficult to differentiate between the two in any given case.

References

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