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Letter to the Editor

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Sneddon's Syndrome: clinical case

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Sir,

We report a 40-year-old female. The high blood pressure was found in Family history, hospitalized in neurology for recurrent ischemic stroke so the etiological assessment was negative, opinion was sought for lesions of livedo at the level of the members back to 2 years without the concept of miscarriage or Raynaud's phenomenon, neither photosensitivity nor a dry syndrome. Dermatologic examination showed dusky *erythematous to violaceous*, irregular, broken circular segments, resulting in a seemingly larger pattern, located on limbs, trunk, and buttocks, exaggerated by the cold and persistent on warming. A report looking for the antiphospholipid antibodies syndrome was positive and the skin biopsy was in favor of a Sneddon syndrome. The patient was treated by anticoagulants and antiplatelet agents with good evolution.

Sneddon's syndrome (SS) is a rare non-inflammatory thrombotic vasculopathy characterized by the combination of Livedo Racemosa (LR) with cerebrovascular disease [1]. Clinico-radiological features of its neurological manifestations, its prognosis and the frequency of associated cardiac valvulopathy remain poorly known, particularly in the absence of Antiphospholipid Antibodies (APL) but it is usually associated with antiphospholipid antibodies [2]. It generally occurs in women between the ages of 20 and 42 years [1,2]. Etiopathogenesis of SS is unknown with 2 primary mechanisms proposed autoimmune/inflammatory versus thrombophilia. The cerebrovascular manifestations are mostly secondary to ischemia (transient ischemic attacks and cerebral infarct). Other neurological symptoms range from headache, cerebral hemorrhage, and early-onset dementia [2,3].

Livedo Racemosa precedes the onset of recurrent strokes by more than 10 years, but in many instances, the significance of the skin lesion is recognized only after the appearance of the stroke. Skin biopsy is critically important for the early diagnosis of SS. Skin biopsy may reveal thrombosis of subcutaneous arterioles and compensatory capillary dilation with blood stagnation causing LR [2]. Although were negative or non-specific in some cases. The main criteria are general LR with typical histopathological findings on skin biopsy and focal neurological deficits [2,4]. Optimal management remains an unresolved problem and long-term anticoagulation has been recommended for ischemic stroke there are controversial results in the treatment of SS with immunomodulatory agents [5].

References

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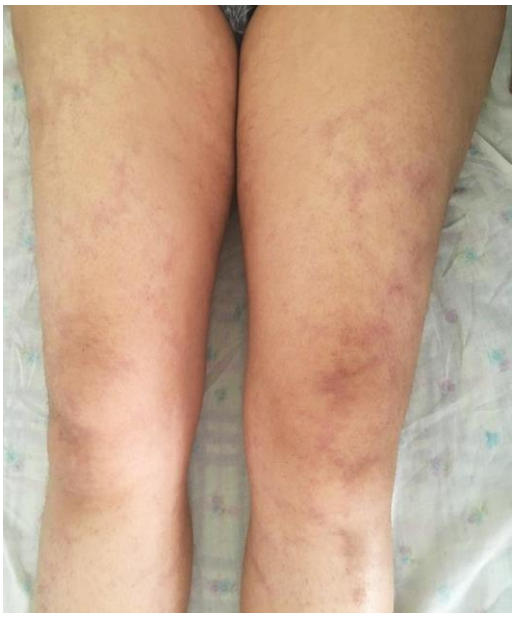
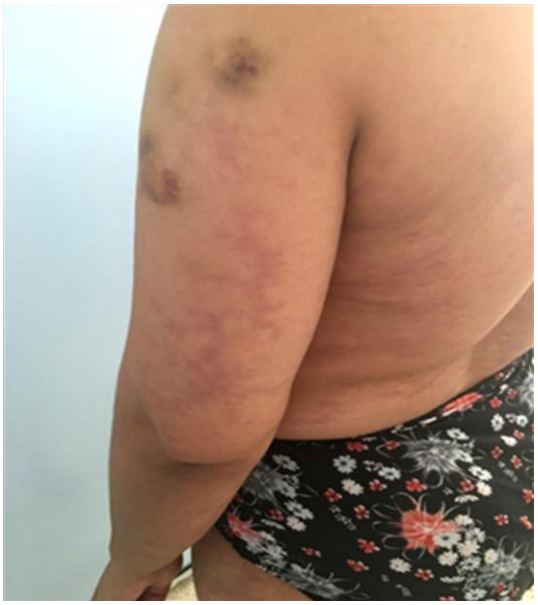
a)



b)



c)

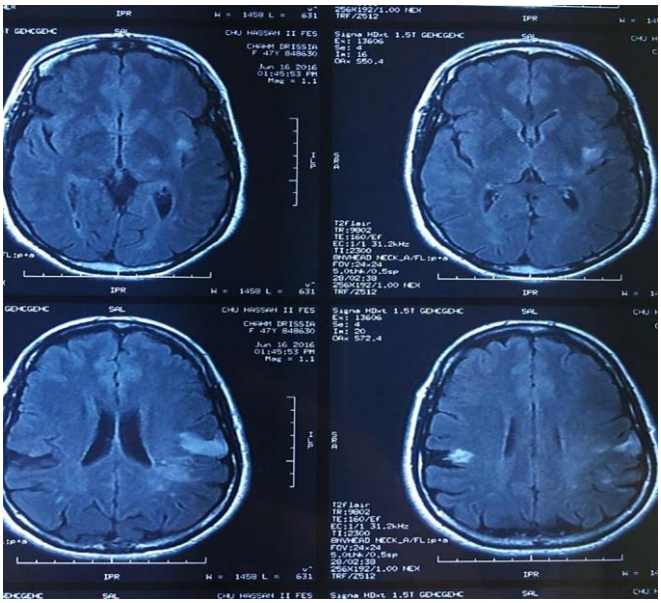


Fig-1:

Livedo racemosa. An erythematous tree branching and forked-lightning pattern on the lower extremities. Please note the broken circle pattern (a,b) . FLAIR MRI brain showing multiple ischemic lesions(c).

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