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Title

A neuro-sweet syndrome mimicking autoimmune encephalitis (case report).

Authors

Ahmed Benmoussa, Samia Khalifa, Amina Chentouf.

Affiliation

Service de Neurologie Médicale - EHU d'Oran Algeria.
Mail: benmoussaahmed79@gmail.com

INTRODUCTION

-Sweet's syndrome, or acute febrile neutrophilic dermatosis, is a sudden onset of painful skin lesions in the form of papules, nodules or asymmetric erythematous plaques in a febrile context associated with a neutrophilic polynuclear infiltrate on skin biopsy.

The Neuro-Sweet syndrome classically presents as encephalitis or febrile meningitis, with skin lesions. In the absence of a preferred cerebral lesion topography, there are no specific signs and a wide variety of symptoms are possible.

AIM

-This is a suspected case of neuro-Sweet syndrome, the clinical picture of which was similar to autoimmune encephalitis.

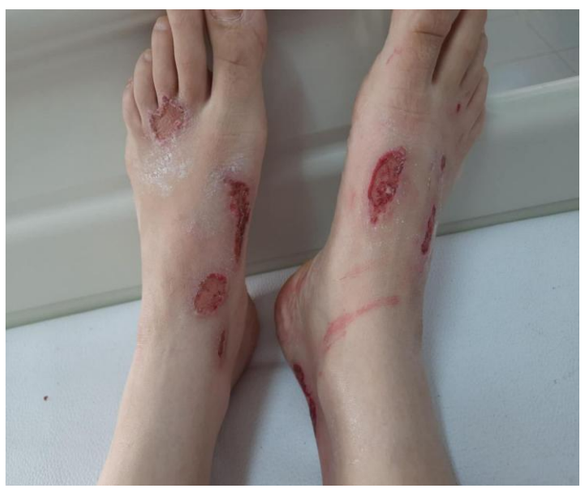
METHODS

- The patient was 26 years old, a married housewife and mother of one child with no particular history of illness. After returning from a trip to Egypt one month later, she presented with an influenza-like syndrome consisting of headache, chills, fever at fever of 38 and asthenia, followed a week later by the onset of an encephalic syndrome with behavioral disorders and left hemicorporeal focal seizures that were secondarily generalized, leading the patient to a state of malaise contemporary with the appearance of painful erythematous pruritic skin lesions with a purplish rim and pigmentation on the lower limbs and abdomen **(photo 01).**

-The patient was transferred to the neurology department after 21 days of TRT with aciclovir and amoxicillin in meningeal doses. She presented with a temporal syndrome with anterograde amnesia, placidity with a feeling of already seen associated with anxiety-type mood disorders with partial motor seizures, and cognitive impairment.

- The white blood cell count was 13,860 ml and the C-reactive protein level was 6.17 mg/l, transaminases were elevated to 3 times normal; TSH was 0.152 iu/ml. Other laboratory tests, including anti-nuclear antibodies, rheumatoid factor, anti-SSA antibodies, anti-SSB antibodies, anticardiolipin antibodies and perinuclear antineutrophil cytoplasm (ANCA), anti NMDA-R Ac were negative, as were HIV serologies, hepatitis C, hepatitis B, EBV, herpes, CBV, West Nile virus and syphilis.

-Lumbar puncture revealed normal cellularity with 2 elements: normoglucorachy, normoproteinorachy.



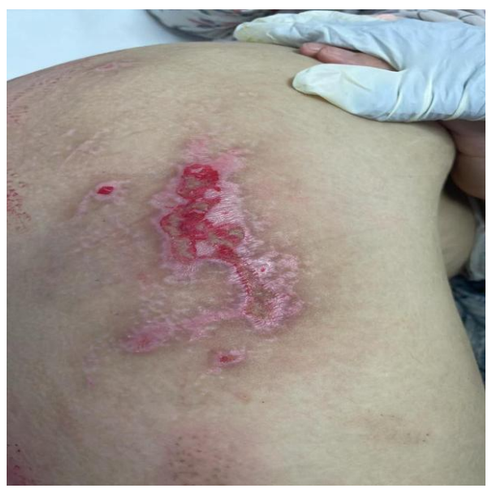


PHOTO 01: erythematous patches on the inside of the feet and the left hip.

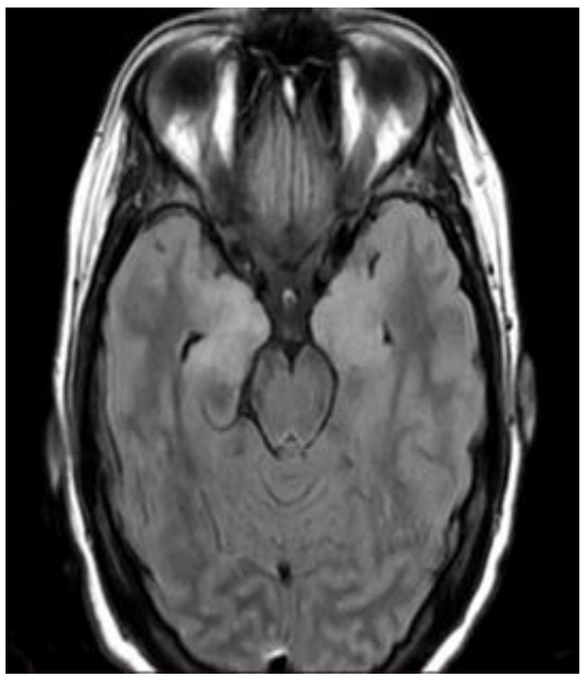


PHOTO 02: Brain MRI Flair sequence showed bilateral medial temporal hypersignal extending to the hippocampal region with areas of diffusion restriction.

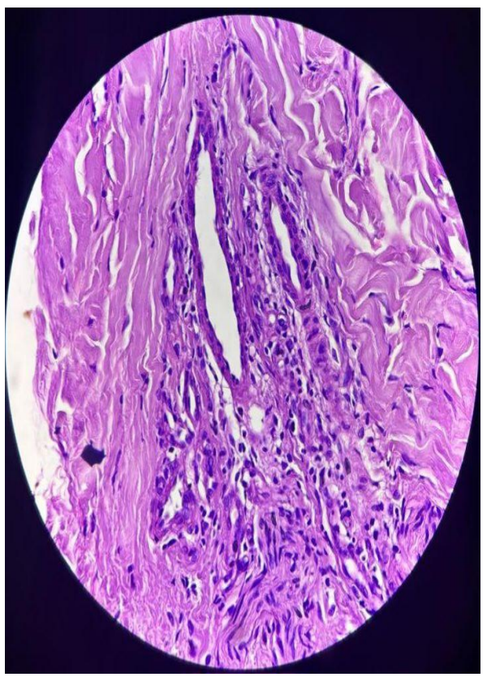


PHOTO 03: perivascular neutrophilic infiltrate

RESULTS

-Brain MRI Flair sequence showed bilateral medial temporal hypersignal extending to the hippocampal region with areas of diffusion restriction **(photo 02)** associated to slow temporo-central spike discharges on EEG; Sweet syndrome was diagnosed on anapathy with neutrophilic infiltrate without leukocytoclastic vasculitis **(photo 03).**

-Progress was favourable with corticosteroid therapy.

DISCUSSION

-The description of the different forms of SS (idiopathic, paraneoplastic and iatrogenic), and the possible association of multiple pathologies, classifies Sweet's syndrome as a systemic pathology. The pathophysiological mechanisms of Sweet's syndrome are poorly elucidated. The presence of IL-1, IL-2 and interferon gamma, but not IL-4, suggests that T1 helper lymphocytes play a role in the pathogenesis of idiopathic variants of this syndrome.

The main differential diagnosis remains neuro-Behcet's syndrome, with similar involvement of the basal ganglia, hyperproteinorachia in the LCR and asymmetric cerebral localisation. The profile of symmetrical limbic encephalitis has never been described.

CONCLUSION

Neuro-Sweet disease should be considered in the differential diagnosis of autoimmune encephalitis; a thorough skin examination and early skin biopsy can facilitate the diagnosis and early treatment of this steroid-sensitive condition.

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