Neuromuscular sweet syndrome: Intermittent relapsing and remitting inflammatory syndrome

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Introduction

Sweet syndrome is a multisystem inflammatory disorder characterized by painful erythematous plaques and aseptic neutrophilic infiltration of various organs including nervous system, muscles, heart, eyes, blood cells and primarily skin. The aetiology of Sweet syndrome is unknown, although the possible role of an abnormally increased chemotaxis of the neutrophils has been suggested. Cytokines, including granulocyte colony-stimulating factor and IL-6 have been implicated in the pathogenesis of Sweet syndrome in few studies. Granulocyte colony-stimulating factor therapy for granulopenia is known to induce Sweet syndrome.[2]

Skin biopsies typically demonstrate dermal infiltration with neutrophils in the absence of vasculitis and it is steroid responsive. There are multiple triggers including infections and drugs. In our case the possible trigger of Sweet syndrome was syphilis in past and the organ systems involved were mainly skin, neuromuscular and haematological.

Case Presentation

This is a case report of a 65-year-old lady who presented with weakness of her left arm and leg, altered GCS , raised creatinine kinase leading to acute renal injury requiring renal replacement therapy. She had few years history of undiagnosed intermittent relapsing and remitting neuro-inflammatory disorder on the background of Myelodysplastic syndrome, neutrophilic dermatosis (sweet disease) and syphilis. Brain imaging performed on this admission did not show any change from her previous scans. While in-patient, she developed generalised tonic clonic seizure episode followed by a rise in Lactate (20), creatinine kinase >70000 and Aspartate aminotransferases >1000 , Type 2 Respiratory failure with persistently low GCS. She then ended up getting intubated and ventilated. No acute changes in MRI apart from longstanding changes and CSF analysis showed pleocytosis and no positive microbiology. Autoimmune encephalitis antibodies were also negative in past and also this time.

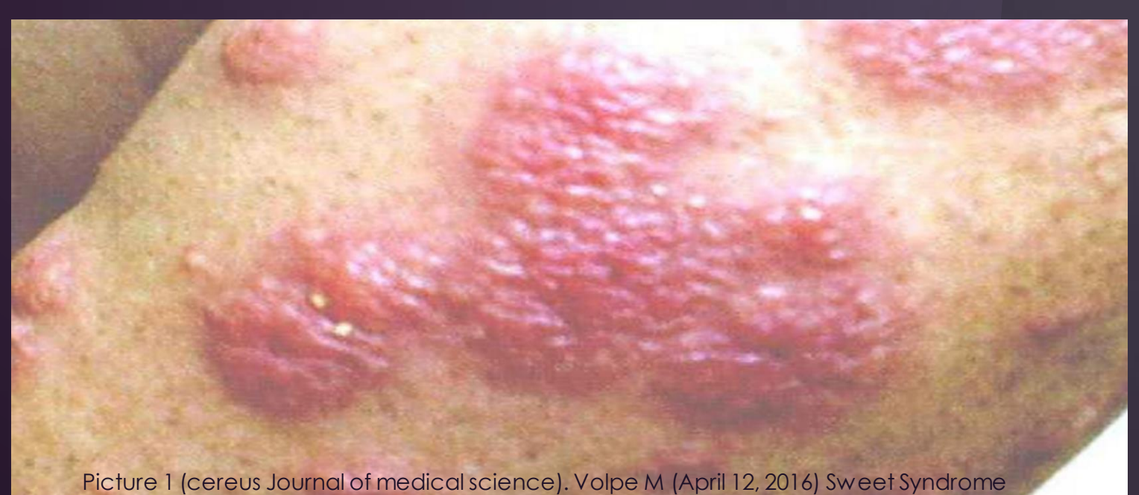
Steroids were commenced in view of possible sweet syndrome and its deterioration with neurological, skin and muscle involvement presenting as weakness, seizures, raised AST, CK and consequently AKI.

After steroids, patient showed remarkable improvement and then later extubated and shifted to ward.

She had similar presentation few years ago when she presented with right sided weakness, seizures, raised AST and CK and was first noted to have MRI changes which showed :

Several areas of subcortical nodular contrast enhancement associated with surrounding vasogenic oedema, within the posterior right frontal lobe and right parietal lobe. Possible similar but non-enhancing signal change within the left posterior frontal lobe."

These MRI changes and her clinical examination findings resolved remarkably on steroids previously as well. Myositis blot and autoimmune profile were all tested negative



Picture 1 (cereus Journal of medical science). Volpe M (April 12, 2016) Sweet Syndrome Associated with Upper Respiratory Infection and Amoxicillin Use. Cureus 8(4): e568. doi:10.7759/cureus.568

No Conflict of Interest

Discussion

In this case, our patient presented with right sided weakness and seizures which were associated with rhabdomyolysis leading to AKI requiring renal replacement therapy. Persistently low Glasgow Coma Score and type 2 Respiratory failure post-ictaly on the background of long history of relapsing remitting steroid responsive neuro-inflammatory disorder led to Intensive care admission. All above mentioned features, biopsy proven neutrophilic dermatosis (skin rash) and Myelodysplastic syndrome comprise parts of sweet syndrome (which encompass multiple organ system including neurological involvement, haematological involvement; with leukaemia's and Myelodysplastic syndrome (as in our patient), primarily skin involvement and associated with infection trigger (syphilis in this case). In terms of muscle involvement with raised *creatinine kinase* and *aspartate aminotransferases* in this case, there have been some case reports of neutrophilic myositis with bullous sweet syndrome.[3]. Steroids remain mainstay of treatment in patients with sweet syndrome / Neuro-muscular Sweet syndrome. We feel in our case having obtained a muscle biopsy (affected organ) to exclude other causes of raised CK and AST including drugs would have consolidated our suspicion of muscle involvement in sweet syndrome although she was remarkably steroid responsive and there was no recent change in drug history, or any other possible explanation besides sweet syndrome.

Conclusions

Sweet syndrome is a rare syndrome and a great mimicker. Although, still a lot to be explored about this syndrome, intermittent relapsing/ remitting nature, neutrophilic inflammatory infiltrates (primarily dermal)involvement of other organ systems and resolution with steroids is pathognomic. All other common causes of similar presentation should be excluded before making the diagnosis

References

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