Atypical presentation of sweet syndrome with oral aphtheform ulceration and abdominal aortic aneurysm

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Abstract

Sweet syndrome is a relatively rare neutrophilic dermatosis characterized by fever, painful skin lesions with systemic involvement. It can occur secondary to multiple underlying aetiological factors. This entity can overlap with other neutrophilic dermatoses such as Behcet's disease.

The patient is a fifty-two-year-old previously well male presented with classic sweet syndrome lesions with systemic unwellness associated with two painful aphtheform ulcerations in buccal mucosa which is relatively a rare presentation of this condition. Further evaluation revealed an incidental finding of an asymptomatic abdominal aortic aneurysm without any significant metabolic risk factors. His pathergy test and HLA B51 were negative. He did not have any other evidence of an underlying condition including haematological or solid organ malignancies, any culprit drugs, recent infections, rheumatoid arthritis or criteria to suggest Behcet's disease. He gave a dramatic improvement for oral steroids and colchicine.

Though he does not fulfil the criteria for Behcet's disease at the moment, being a well-known entity which overlaps with sweet syndrome, these two features of aphtheform oral ulcerations and vascular involvement in the form of abdominal aortic aneurysm could be part of evolving Behcet's disease and he should be longitudinally followed up for further development of symptoms.

Introduction

Sweet syndrome is a rare multisystemic inflammatory disorder with combination of fever, neutrophilic leukocytosis, painful erythematous cutaneous plaques with dermal neutrophilic infiltration. Multiple underlying aetiological associations are described in the literature including malignancy, drugs, infections, and other neutrophilic dermatoses including Behcet's disease. It has a dramatic response to systemic corticosteroids.

Case report

Fifty-two-year-old previously healthy male presented with one-week history of fever and systemic unwellness associated with 3 days history of painful erythematous skin nodules and plaques (Figure 1,2) with pseudo vesicular appearance distributed over head and neck area, dorsum of hands and few over legs. He also complained of two painful aphtheform ulcerations in buccal mucosa (Figure 3). He denied any history of recurrent oro-genital ulcerations, ocular involvement or other features suggesting Behcet's disease. No significant underlying aetiologies were found in further evaluation. His investigations revealed neutrophil leucocytosis with high inflammatory markers and dermal infiltration of neutrophils in skin biopsy suggestive of sweet syndrome. Malignancy screening revealed a normal blood picture and serum protein electrophoresis with normal upper and lower gastro intestinal endoscopies. His contrast abdominal CT scan revealed an incidental finding of abdominal aortic aneurysm (4.8cm AP/4.4cm Tr) with no evidence of leaking or rupture. His pathergy test and HLA B51 antigen were negative.

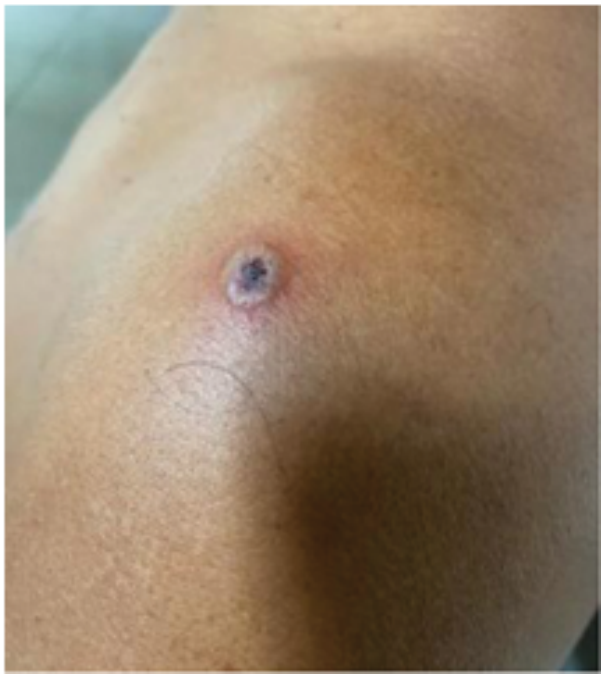

Figure 1.

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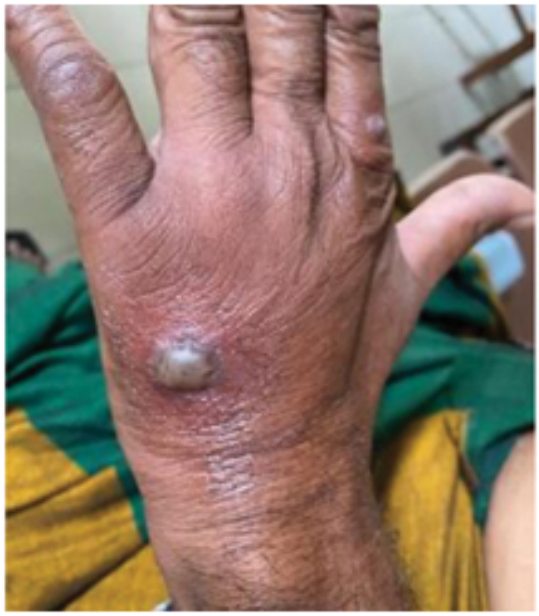

Figure 2.

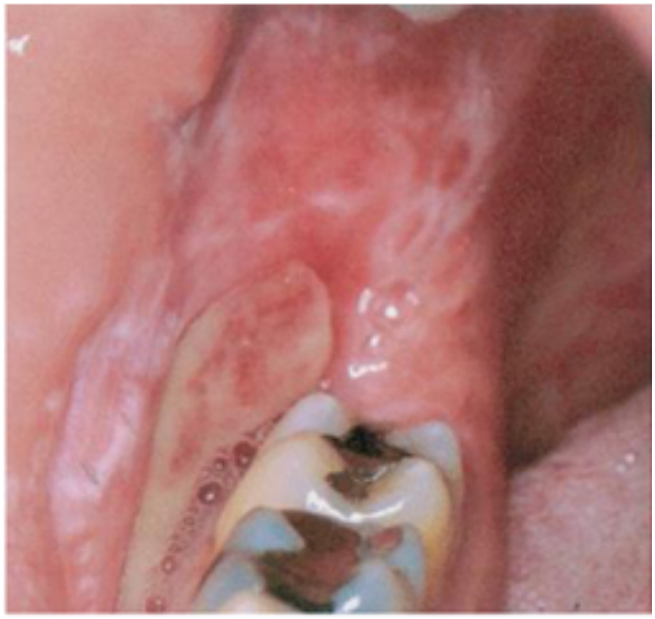

Figure 3.

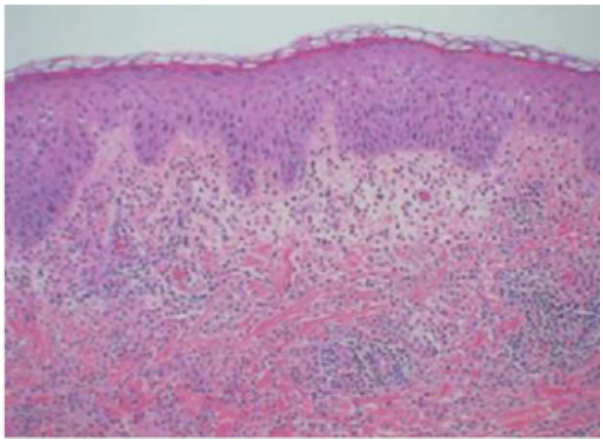

Figure 4.

He was started on oral prednisolone 60mg daily with colchicine 0.5mg tds for which he showed a dramatic response. A vascular surgical opinion was taken with regard to aortic aneurysm and decided on conservative management.

Discussion

Although this patient doesn't fulfil the criteria for Behcet's disease it is well known for both entities to occur as an overlap condition. In classic sweet syndrome it is rare to have aphtheform oral ulcerations and abdominal aortic aneurysm would be a feature of vascular Behcet's. Therefore it is important to have a longitudinal follow-up for further development of skin lesions and features of Behcet's disease in this patient.

References

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