

Sweet Syndrome Revealing Systemic Lupus Erythematosus

Abstract:

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Abstract

Sweet Syndrome is an acute inflammatory skin eruption which is rare in children. We report a case of childhood Systemic Lupus Erythematosus (SLE) that presented with Sweet syndrome. This case is a unique presentation of a common disorder which provides a new facet for the differential diagnosis of SLE in children. It is also the first paediatric case to be reported in a Caucasian child.

Introduction

SLE is a chronic auto-immune disorder characterised by multi-system organ involvement and marked clinical heterogeneity. Although the underlying aetiology remains to be fully elucidated, it is thought that SLE may be triggered by several factors including environmental pathogens and infection in those with an underlying genetic predisposition¹. Sweet syndrome is a reactive neutrophilic dermatosis which is rare in children². It was described in 1964 by Dr Robert Douglas-Sweet, who documented an acute inflammatory skin eruption associated with fever and leucocytosis³. Diagnosis requires the presence of two major and two of four minor criteria⁴. Major criteria include abrupt onset of painful erythematous plaques/nodules and histological evidence of dense neutrophilic infiltrate. Minor criteria include pyrexia >38°C; elevated ESR/CRP/leucocytes/neutrophils; response to corticosteroids and association with an underlying haematological disorder, inflammatory disease or recent respiratory or gastrointestinal infection⁵. Characteristically, Sweet-syndrome responds rapidly to corticosteroids⁶.

Case Report

A 12 year old female developed generalised arthralgia, lethargy and anorexia 3 weeks following an upper respiratory tract infection. She was systemically unwell with pallor, drowsiness and dehydration. She developed swelling and restriction of the right wrist and left ankle. Examination of the skin revealed a papulo-vesicular rash on the elbows and ankles (Figure 1). Initial investigations showed anaemia: (haemoglobin:11.1g/dL) with thrombocytosis(platelets:467x10⁹/L). ESR and CRP were raised (99mm/hr and 84mg/l respectively). Liver transaminases were elevated: AST=192U/l, ALT=596U/l. Serum albumin was low: 27U/l. Complement C4 was reduced: 0.07g/L while immunoglobulin levels revealed high IgG:57.3g/L. Biopsy of the papulo-pustular lesion revealed a florid neutrophilic infiltrate with apoptotic debris smeared between collagen, confirming severe neutrophilic dermatosis and a diagnosis of Sweet-syndrome (Figure 2). The differential diagnosis included SLE, malignant haematological disease and gastroenterological disease. The remainder of the immunological profile demonstrated a strongly positive homogenous ANA (1:2560) and anti-DS-DNA antibodies were evident at 26 IU/ml. Anti-cardiolipin IgG antibodies were elevated at a titre of 41 GPLU/ml. She was commenced on intravenous methyl-prednisolone to which she responded well. Subsequent to this admission, the DNA crithidia titre was found to be significantly elevated: el:160 and DNA-ELISA was elevated at 16 IU/ml confirming SLE. She was commenced on azathioprine. At last review she had no joint pain or swelling and the rash had completely resolved.

Discussion

Recent studies suggest that childhood-onset SLE has a more severe disease course than adults. Aggressive treatments incorporating biological agents have improved survival in childhood SLE, but the disease is still associated with significant morbidity. Early diagnosis is essential if disease-associated complications are to be prevented. The association of SLE with Sweet syndrome has been reported in adults⁷, but to date there is only 1 report in the paediatric population⁸. The pathogenesis of Sweet syndrome remains incompletely understood. It has been proposed that it may result from a hypersensitivity reaction to an antigen such as bacteria, virus or even tumour. Different studies have implicated the roles of immune complexes, autoantibodies, cytokines, dermal dendrocytes, HLA serotypes and leucotactic mechanisms in its development⁹. It has also been postulated that photosensitivity plays a role in its development providing a possible link with photosensitivity commonly associated with SLE. There are very few reports of Sweet syndrome in paediatric patients. One of the youngest patients reported was 5 weeks of age and subsequently found to have non-B54 HLA types¹⁰. Despite this there is no clear genetic association. Another case reports an infant with known chronic granulomatous disease who presented with methicillin sensitive staphylococcus aureus lymphadenitis supporting the hypothesis that Sweet syndrome may be part of a complex of inflammatory conditions. Sweet syndrome has been associated with a variety of disease states including acute myeloid leukaemia and inflammatory bowel disease. Newer treatment options include intravenous immunoglobulin and anti-interleukin-1 receptor antagonists¹¹.

Sweet syndrome is rare but its onset may herald a serious underlying disorder requiring prompt diagnosis and treatment. This case reports an unusual presentation of SLE, which highlights the importance of considering it in the differential diagnosis of a child with Sweet syndrome. The child in this report was treated early with excellent outcome, emphasising the importance of recognizing and treating early this emerging disease association.

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