Sweet’s Syndrome with Tendinitis

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Abstract

A 30 years man presented with fever and skin manifestations in the form of multiple tender, erythematous, asymmetric nodules and papules predominantly over the upper arms and back. The nodules were coalescing to form irregular sharply demarcated plaques over the forearms and thighs. There were pustules over the nodules. He also had tendoachilles tendinitis. The total leucocyte count, differential count and skin biopsy confirmed the diagnosis of Sweet’s syndrome (acute febrile neutrophilic dermatosis). This patient, a case of Idiopathic Sweet’s syndrome with tendoachilles tendinitis responded to oral corticosteroids.

INTRODUCTION

Acute febrile neutrophilic dermatosis, a clinical and histologically well delineated entity, which was thought to be rare has about 500 cases reported in literature. The pride of discovery of this condition goes to the English dermatologist, Robert Douglas Sweet from Plymouth in 1964 who described this condition in eight female patients.

We present a case of acute febrile neutrophilic dermatosis which is well known for its female preponderance, in a male aged 30 years, with no predisposing risk factors. The striking feature in this patient was the presence of Achilles tendinitis, a rare manifestation not hitherto reported in literature.

CASE REPORT

This 30 years man, presented with a history of irregular fever for three weeks and malaise, joint pain, heel pain and multiple painful skin lesions for one week. He did not complain of loss of appetite or weight. He had no symptoms pertaining to the eye, gastrointestinal, respiratory, genito-urinary tracts or the nervous system. He had no previous similar episodes or vaccination and had not taken any drugs except paracetamol.

On examination he was mildly febrile and anaemic. There was no significant lymphadenopathy. The oral mucosa, nails, palms and soles were normal.

He had multiple tender, erythematous, asymmetric nodules and papules over the arms [Fig. 1] and back. There were pustules over the nodules [Fig. 1]. The nodules were coalescing to form irregular sharply demarcated plaques over the forearms and thighs. Musculoskeletal examination revealed right Achilles tendinitis [Fig. 2].

The investigations revealed – hemoglobin [Hb] 12g%, erythrocyte sedimentation rate [ESR] by Westergren method of 110mm at the end of one hour, total leucocyte count [TLC] 12,800 cells/cumm, differential count [DC] polymorphs 73, lymphocytes 24, eosinophils 3, platelets 2.83 lakhs/cu.mm with a normal peripheral smear and bone marrow study. Urine examination was normal. ELISA for HIV and Mantoux were negative. The blood Widal, bacterial culture of throat swab, blood, urine and stool were negative. Biochemical parameters like blood sugar, urea, creatinine, SGOT, SGPT and serum alkaline phosphate [SAP] which was 12.9 KA units were normal. The C-reactive-protein was 24mg/l, antistreptolysin-O, rheumatoid factor, antinuclear antibody [ANA] and anti neutrophil cytoplasmic antibody [ANCA] were negative. X-ray chest and ultrasonogram of the abdomen were normal. Ultrasonogram of the right Achilles tendon at its insertion revealed increased width with more hypoechoegenecity suggestive of inflammation [Fig. 4].

Biopsy of the skin revealed papillary dermal edema, with a dense perivascular inflammatory infiltrate in the upper and mid dermis consisting of neutrophils and a few lymphocytes. Swollen endothelial cells were present [Figs. 3]. There was no evidence of vasculitis.

He was treated with 40mg of prednisolone per day which was tapered over a period of four weeks. The skin lesions resolved and pain, swelling and tenderness over the Achilles tendon reduced markedly.

DISCUSSION

Sweet’s syndrome is characterized by the sudden onset of fever, leucocytosis, tender, erythematous well demarcated papules and plaques favouring mostly the face and upper part of the body occasionally being pseudovesicular, pseudopustular, vesicles, pustules and
bulla. Akin to HIV, syphilis and systemic lupus erythematosus (SLE), multisystem including musculoskeletal involvement is common.³

Although the exact etiopathogenesis of Sweet’s syndrome is not known, it may be a hypersensitivity reaction associated with elevated cytokine levels like IL-1, IL-6, IL-8, G-CSF and GM-CSF. HLA BWS4 association has been noted in Japanese patients.⁵

Von Den Driesch in 1994² suggested that Sweet’s syndrome may be subdivided into four groups namely classic idiopathic, parainflammatory, paraneoplastic and pregnancy associated. Majority of the cases are idiopathic constituting 71% with 11-54% secondary to hematological malignancies and 19% being associated with immunological disorders like relapsing
polychondritis, rheumatoid arthritis, dermatomyositis and also inflammatory bowel disease in the 190 female cases in Mayo clinic studies.5 Other rheumatological associations include SLE, Sjogren’s syndrome, sarcoidosis and Behcet’s disease. Our patient did not have clinical or investigational evidence for rheumatological disease and malignancy.

The idiopathic form has a female preponderance with a female to male ratio of 3:1, though in our case it was a male.2 To the best of our knowledge till date there have been no reports of idiopathic Sweet’s syndrome patients with Achilles tendinitis, although arthralgia and arthritis have been observed in 33-62% of patients, with pauciarticular arthritis in 25%. A single case report of recurrent tenosynovitis in a 50 year old woman has been reported.6

As per the diagnostic criteria the white blood cell count should be higher than 8000 cells/cumm with polymorphs constituting more than 70%.2 Serum alkaline phosphatase which is found to be elevated in 46% of patients was normal in our patient. In this patient ANCA was negative, as has been in recent reports.

The characteristic histopathological features are a dense dermal perivascular inflammatory infiltrate and dermal edema.2 The typical features of acute febrile neutrophilic dermatosis in the skin biopsy of this patient ruled out the possibilities of erythema multiforme, erythema nodosum, pyoderma gangrenosum, erythema elevatum diutinum and skin lesions of SLE which are mimics of this disease.3,4

This patient responded dramatically to steroids. The other drugs that have been tried are indomethacin, potassium iodide, colchicine, cyclosporine, chlorambucil, cyclophosphamide, sulphapyridine, clofazamine and IFN α2. 25% of cases develop recurrence.

Idiopathic Sweet’s syndrome in a 30 years male with predominant skin manifestations over the upper torso, arthralgia and an unusual presentation like Achilles tendinitis were the highlighting features in this patient.

REFERENCES