AN ATYPICAL PRESENTATION OF SWEET’S SYNDROME WITH AN UNUSUAL AND EXAGGERATED PATHERGY: A CASE REPORT

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ABSTRACT

Sweet’s syndrome (SS) is a rare disorder first described by Sweet in 1964. [1] It is a neutrophilic dermatosis. It manifests with acute onset of high grade fever, neutrophilia, tender erythematous papules, nodules, and plaques, and histopathology characterized by a diffuse infiltrate consisting predominantly of mature neutrophils that are typically located in the upper dermis. We report a case of atypical presentation of idiopathic sweet’s syndrome with an unusual and exaggerated pathergy phenomenon.

KEYWORDS

Neutrophilic Dermatosis, Idiopathic Sweet’s Syndrome, Unusual and Exaggerated Pathergy Phenomenon.

INTRODUCTION

This is a case of sweet’s syndrome with an unusual presentation. This case also illustrates an association of the pathergy phenomenon with Sweet’s syndrome. Sweet’s syndrome is a rare disease as only over 500 cases of sweet’s syndrome have been reported worldwide. [2] In literature, pathergy is rarely reported in those with Sweet’s syndrome, but it is known to occur in up to 8% of patients. [3] In this case there is an exaggerated and unusual pathergy phenomenon.

KEY MESSAGES

What was known?
Pathergy in Sweet’s syndrome patients is rarely reported. [2] The Pathergy test is the skin hyper reactivity associated with erythema, papules, or pustules which is induced by intradermal prick. [10]

What is New?
Sweet’s syndrome can present with a longer duration. The pathergy phenomenon can manifest in the form of bullae and ulcerations and the pathergic response can be exaggerated, spreading beyond the site of trauma.

CASE HISTORY

A 35 years - old male, presented with a 3-month history of fever, arthralgia of both knee joints and both elbow joints, myalgia and malaise. There was a history of preceding minor trauma. On examination he had multiple, tender, irregular erythematous oedematous plaques, with local rise of temperature and a few hyperpigmented papules on the left lower limb [Figure 1, 2]. His systemic examination was normal. He had pallor and left inguinal lymph nodes were enlarged. The oral and genital mucousae were normal.

The lab investigations revealed Anaemia with Haemoglobin% 10gm%, Leucocytosis with total leucocyte count 8800 cells/mm³ and a neutrophilic predominance of 72%, elevated C-reactive protein and Erythrocyte sedimentation rate. Peripheral smear showed mild degree normocytic hypochromic anaemia, FNAC from left inguinal lymph node showed Nonspecific lymphadenitis. His Urine examination, Liver function tests, Renal function tests, Chest X-ray (PA view) and Ultrasound Abdomen were normal, Sputum for acid fast bacilli was negative and ASO titres were not raised.

Skin biopsy showed mild focal spongiosis in the epidermis. There is a diffuse dense infiltrate of neutrophils interspersed with nuclear dust and lymphocytes involving the whole upper and mid dermis. The papillary dermis is oedematous several small vessels in mid and lower dermis show fibrin deposition within their walls along with leucocytoclasia. These features are suggestive of Sweet’s syndrome [Figure 8].

As a routine screening for tuberculosis Mantoux test was performed and the patient was started on IV antibiotics. He developed oedema, vesicles and bullae at the site of IV cannula, Mantoux test and skin biopsy [Figure 3 - 7]. The lesions at the site of Mantoux test and skin biopsy ulcerated with a surrounding oedema suggesting a positive and exaggerated pathergy phenomenon [Figure 3, 4, 6, 7]. We diagnosed the case as Sweet’s syndrome as the diagnostic criteria for Sweet’s syndrome was fulfilled. We started the patient on oral prednisolone 60mg/day. The dose of steroid was tapered every week. The skin lesions cleared rapidly with post inflammatory hyperpigmentation [Figure 8, 9] and the patient was on follow up.

![Fig. 1: Image showing edematous plaques and hyper pigmented papules](image-url)
Fig. 2: Edematous plaques and hyperpigmented papules over left lower limb

Fig. 3: Lesion at the site of mantoux test showing ulceration and edema indicating an unusual and exaggerated pathergy

Fig. 4: Lesion at the site of Mantoux test showing ulceration and oedema extending beyond the lesion

Fig. 5: Oedematous plaque with pseudovesiculation at site of IV cannula and papules extending beyond the site of IV catheter

Fig. 6: Site of skin biopsy developing bullae, ulcers and oedema extending beyond the site of skin biopsy

Fig. 7: Site of biopsy developing blisters, ulcers and oedema extending beyond the lesions

Fig. 8 (a): Histopathology viewed under 10 X magnification with H & E staining, showing neutrophilic infiltration and dermal oedema

Fig. 8 (b): Histopathology viewed under 40 X magnification with H & E staining, showing neutrophilic infiltration and dermal oedema
Fig. 9: After treatment with systemic steroids complete resolution of lesions with some post inflammatory hyperpigmentation

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<td>Abrupt onset of painful erythematous plaques or nodules</td>
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<td>Histopathologic evidence of a dense neutrophilic infiltrate without evidence of leukocytoclastic vasculitis</td>
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<th>Minor Criteria</th>
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<td>Pyrexia &gt;38°C</td>
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<td>Association with an underlying hematologic or visceral malignancy, inflammatory disease, or pregnancy, or preceded by an upper respiratory or gastrointestinal infection or vaccination</td>
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<td>Excellent response to treatment with systemic corticosteroids or potassium iodide.</td>
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<td>Abnormal laboratory values at presentation (Three of four): Erythrocyte sedimentation rate &gt;20 mm/hr; positive C-reactive protein; &gt;8,000 leukocytes; &gt;70% neutrophils.</td>
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<th>Table 1: Diagnostic Criteria for Sweet’s Syndrome</th>
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<td>Both major criteria and two of the four minor criteria are required to establish the diagnosis of classical SS.</td>
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DISCUSSION

Sweet’s syndrome is an acute febrile neutrophilic dermatosis. The diagnostic criteria for classical Sweet’s syndrome were originally proposed by Su and Liu[7] in 1986. They were modified by von den Driesch [9, Table 1] in 1994. Both major criteria and two of the four minor criteria are required to establish the diagnosis of classical SS.

Our patient met all, but one minor criterion and radiological, biochemical and haematological investigations did not reveal any underlying malignancy or any systemic association. Hence this is a case of classical idiopathic Sweet’s syndrome. Classical or idiopathic Sweet’s syndrome typically affects women in the third to fifth decade. [4] The cutaneous manifestations consist of erythematous to violaceous tender plaques. The lesions have a transparent, vesicle-like appearance secondary to the pronounced oedema in the upper dermis (Pseudo vesicular appearance). [4] The lesions typically involve the arms, face and neck, but may occur anywhere, and are usually multiple but may be solitary.

Our patient is a 35-year-old male who presented with tender, indurated, oedematous, erythematous plaques on the lower limbs and a few hyperpigmented papules on the left thigh. They gradually progressed over a period of 3 months to involve the left leg up to the ankle joint. Pathergy is defined as an induction of dermatosis-associated skin lesions appearing at sites of skin trauma. [2] There have been reports of a positive skin pathergy at the sites where procedures such as biopsies, [2] intravenous catheter placement, [2] and venepuncture have been performed. [2] In our patient oedema and blisters appeared at the sites of intravenous catheter, Mantoux test and skin biopsy. Later there was ulceration with a surrounding oedema at these sites suggesting an exaggerated and unusual pathergy phenomenon.

The diagnostic histopathological features of Sweet’s syndrome include a dense, predominantly neutrophilic infiltrate located in the superficial dermis, and prominent papillary dermal oedema which may occasionally lead to subepidermal vesication. Lymphocytes, eosinophils and histiocytes may be present. The infiltrate often occurs in a diffuse pattern, but may be perivascular or have an upper dermal band – like distribution Neutrophil karyorrhexis (Fragmented neutrophil nuclei; nuclear dust) is a common finding. The epidermis is often normal, but spongiosis may be present, and rarely neutrophils may extend into the epidermis to form subcorneal pustules. The skin biopsy specimen of our patient has shown the characteristic features of Sweet’s syndrome.

Sweet’s syndrome is known to respond dramatically to oral steroids. Second-line therapies include potassium iodide, colchicine, indomethacin, dapsone, clofazimine, doxycycline and metronidazole. [11] We treated the patient with a tapering course of systemic steroids. The lesions healed rapidly without scarring. In this case although Sweet’s syndrome is an acute neutrophilic dermatosis, the patient had a history of fever and arthralgia for 3 months during which the lesions gradually increased in number and size. In addition to this the patient had plaques and papules as the primary lesions while the pathergy was unusual and exaggerated and manifested as erythema, oedema and bullae that ruptured to form erosions and ulcers with a surrounding oedema which spread beyond the sites of trauma.

REFERENCES