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Case Report

Eosinophilic Dermatosis of Hematological Malignancy Mimicking Wells Syndrome

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Abstract

Wells syndrome, also known as eosinophilic cellulitis, is an inflammatory dermatitis that is often misdiagnosed as infectious cellulitis, leading to the inappropriate use of antibiotics and a delay appropriate treatment. We present a diagnostically challenging case where a 65-year-old male with a known case of SLL was found to have eosinophilic dermatosis of hematological malignancy after a thorough workup. Although the association between CLL and Wells syndrome is well established, we have not encountered any similar association or other cases of Wells syndrome associated with SLL in the literature. The pathophysiology underlying the association between eosinophilic cellulitis and CLL/SLL remains incompletely understood, and further investigation is warranted.

Keywords: Wells Syndrome; Small Lymphocytic Lymphoma; Eosinophilic Dermatosis of Hematological Malignancy; Eosinophilic Cellulitis

Introduction

Wells syndrome, also known as eosinophilic cellulitis, is an inflammatory dermatitis that typically presents with erythematous plaques, although the presentation can vary significantly. Eosinophilic cellulitis, as the name implies, can present similarly to erysipelas/cellulitis and is often misdiagnosed as infectious cellulitis [1]. Early accurate identification is key, as misdiagnosis can lead to the inappropriate use of antibiotics and delay appropriate treatment. Thus, we present a case of a 65-year-old male with a known case of small lymphocytic lymphoma (SLL) who developed eosinophilic dermatosis of hematological malignancy (EDHM).

Case Presentation

A 65-year-old Bahraini male with a known case of small lymphocytic lymphoma (SLL) since 2015, which was being followed by hematology, presented to the accident and emergency in May 2020 with one week of itchy skin lesions on his face and arm and swelling of the foot. He also reported that over the last year, he has had on/off lesions of similar character distributed diffusely on his body associated with swelling of the limbs. He was initially believed to have an insect bite with cellulitis and was prescribed several courses of antibiotics, although he denied any recollection of a preceding insect bite. After resistance to antibiotic treatment, he had a skin biopsy and was then incorrectly diagnosed as having a drug reaction. On physical examination, the patient was well-appearing and a febrile. He had an erythematous plaque with multiple small pustules in the periphery on his forehead. In addition, there were multiple erythematous violaceous papules, nodules, and plaques on his elbow, left leg and right foot with significant swelling, redness and change in the temperature (Figure 1A-1D). The initial differential diagnosis included: wells syndrome, erythema elevatum diutinum, sweet syndrome, and leukemia cutis.

His labs revealed leukocytosis with lymphocytosis and a normal eosinophil count. Skin biopsy from the elbow lesion showed intact epidermis with distinct intraepidermal/suprabasal bullae formation with marked papillary edema and associated predominant eosinophils infiltration in the underlying dermis (perivascular/peri-appendageal distribution) and subcutaneous fat focally. There was no evidence of any necrotizing vasculitis or atypically pathology present. Immunohistochemical staining was CD3 positive, Bcl-2 focally positive, CD20 positive in rare cells, and CD10 negative, excluding leukemia cutis (Figure 2A-2C). These findings were consistent with Wells disease-bullous type. All other labs were within normal ranges, including stool testing for parasites.

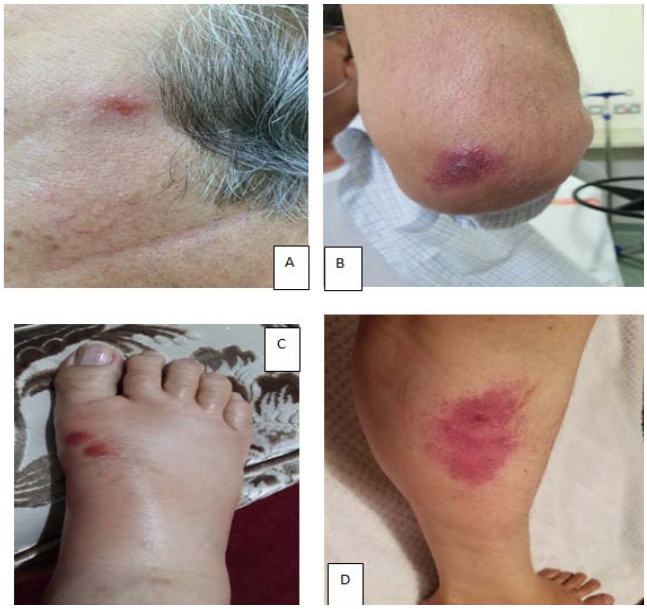

Figure 1: A) Erythematous plaque with multiple small pustules in the periphery on forehead. B-D) Multiple erythematous violaceous papules, nodules, and plaques on his elbow, left leg and right foot with significant swelling.

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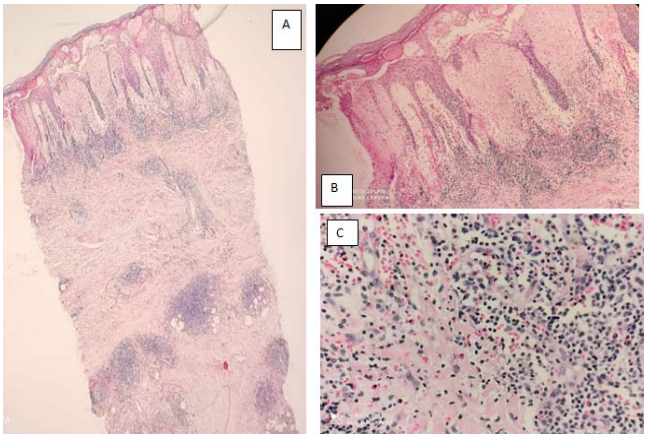

Figure 2: Skin biopsy of elbow lesion: A) At 10x-magnification: Epidermis with distinct intraepidermal/supra-basal bullae formation with marked papillary edema and associated predominant eosinophils infiltration in the underlying dermis. Perivascular/peri-appendageal distribution including the subcutaneous fat focally. B) At 20x-magnification: Intraepidermal supra-basal bulla formation with marked papillary edema. C) At 40x-magnification: Predominant eosinophils infiltration in the underlying dermis.

Our impression based on both the clinical and histological finding was consistent with eosinophilic dermatosis of hematologic malignancy. The patient was treated with systemic steroids and an antihistamine drug, and his symptoms improved dramatically. However, the condition recurred within 2 months. In addition, the case was discussed with his hematologist for further management of his SLL, as his skin findings suggested the possible progression of his hematological condition.

Discussion

Eosinophilic cellulitis is a rare inflammatory dermatosis first described by Dr. Wells that is characterized by recurrent granulomatous dermatitis with eosinophilia [2]. Although the etiology of Wells syndrome is unknown, it is hypothesized that the condition may be a manifestation of an autoimmune reaction and is mediated by a Type IV hypersensitivity reaction [3,4]. Eosinophilic cellulitis typically presents with a constellation of nonspecific symptoms including rash, fever, and arthralgia, and patients most commonly report pruritus as a symptom [5,6]. Skin manifestations can vary from erythematous plaques to pruritic vesicles and/or bullae, which can contribute to misdiagnosis as cellulitis/erysipelas [5]. Wells syndrome has been associated with a number of precipitating factors, including insect bites, viral infections, and drug reactions [7]. Other conditions believed to trigger eosinophilic cellulitis include hematologic and oncologic disorders, such as chronic lymphocytic leukemia (CLL), non-Hodgkin's lymphoma, and hypereosinophilic syndrome.1 However, SLL, a variation of CLL affecting primarily B-cells of the lymph nodes, has not been previously associated with Wells syndrome [8].

Wells syndrome, particularly when presenting with an underlying lymphoma, can be diagnostically challenging. Wells syndrome, first described by Weed in 1965, was initially thought to be due to an exaggerated or hypersensitivity reaction to insect bites in patients with CLL, but patients were unable to recall insect bites preceding the dermatosis [9]. The connection of Wells syndrome with underlying hematological malignancies was later established. Thus, Wells syndrome has also been referred to as eosinophilic dermatosis of hematologic malignancy, given the vast spectrum of hematologic conditions that are capable of precipitating the skin manifestations [10]. In addition, the polymorphic cutaneous presentation of eosinophilic dermatitis of hematologic malignancy can resemble and be diagnosed as cellulitis/erysipelas, which will be resistant to treatment with antibiotics and require reconsideration of the differential diagnosis. This was true in this case, as the patient was incorrectly diagnosed with and/or treated for cellulitis and insect bites before arriving to the correct diagnosis. When treating patients with SLL/CLL who develop cutaneous lesions, especially when due to a suspected severe arthropod reaction or refractory cellulitis, physicians should maintain a reasonable suspicion of eosinophilic dermatosis of hematological malignancy in the differential diagnosis.

Although the association between CLL and Wells syndrome is well established, we have not encountered any similar association or other cases of Wells syndrome associated with SLL in the literature. However, given that SLL represents a varying presentation of CLL, we suspect that much of what is known in regards to diagnosing and treating CLL with Wells syndrome may hold true for patients with SLL. Despite the similarities of CLL and SLL, it remains unclear as to what differences may exist in the predisposition for developing Wells syndrome and the impact on outcomes, severity, etc. The pathophysiology underlying the association between eosinophilic cellulitis and CLL/SLL remains incompletely understood, and further investigation is warranted.

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