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BULLOUS WELLS SYNDROME: A CASE REPORT.

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ABSTRACT

Bullous Wells syndrome is a rare inflammatory dermatosis. Typically it presents as an erythematous plaque with bullae. Morphologically, it can be confused with bullous cellulitis, arthropod bite, contact dermatitis and bullous pemphigoid. Histopathology is mandatory to diagnose the disease. Bullous Wells syndrome can be associated with sinister systemic conditions such as Churg Strauss syndrome, ulcerative colitis and lymphoma. Treatment can be topical or systemic steroid. We report an interesting case of idiopathic bullous Wells syndrome in a 30-year-old woman who responded well to oral steroid.

Keywords: **Bullous Lesions, Drug Therapy, Eosinophilia, Eosinophilic Cellulitis, Wells syndrome.**

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ABSTRACT
Bullous Wells syndrome is a rare inflammatory dermatosis. Typically it presents as an erythematous plaque with bullae. Morphologically, it can be confused with bullous cellulitis, arthropod bite, contact dermatitis and bullous pemphigoid. Histopathology is mandatory to diagnose the disease. Bullous Wells syndrome can be associated with sinister systemic conditions such as Churg Strauss syndrome, ulcerative colitis and lymphoma. Treatment can be topical or systemic steroid. We report an interesting case of idiopathic bullous Wells syndrome in a 30-year-old woman who responded well to oral steroid.

Keywords: **Bullous Lesions, Drug Therapy, Eosinophilia, Eosinophilic Cellulitis, Wells syndrome.**

INTRODUCTION
Bullous eosinophilic cellulitis or Wells syndrome is an uncommon inflammatory disorder.[1, 2] It typically presents as an oedematous and erythematous plaque, with presence of bullae. Clinically, it can mimic bullous cellulitis, intense arthropod bite, bullous pemphigoid or intense contact dermatitis. Diagnosis is based on history, examination, blood investigation and histopathological findings. Peripheral blood may reveal eosinophilia in approximately 50% of cases.[1] Histological features include diffuse dermal eosinophil infiltration with intradermal or subepidermal blisters. The most characteristic finding is flame figures.

Bullous Wells syndrome has been reported to be associated with more sinister systemic conditions such as ulcerative colitis, lymphoma and Churg-Strauss syndrome.[3-6] Although it can resolve spontaneously, Wells syndrome typically follows a relapsing and remitting pattern.[1, 2, 6] Treatment involves topical or systemic steroid, as well as treating the associated condition if present. The purpose of this case report is to highlight the importance to consider bullous Wells syndrome in a patient presenting with blistering dermatoses. Failure to diagnose may lead to unnecessary treatment and increased morbidity.

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CASE REPORT

A 30-year-old woman presented with red, pruritic patches and plaques on bilateral legs for three-week duration. She had a background history of atopic dermatitis. Physical examination revealed she was febrile. There were tense blisters filled with serosanguineous fluid on the dorsal aspect of both feet (Figure 1). No signs of skin trauma, insect bite or regional lymphadenopathy.

Investigations showed leukocytosis with eosinophilia. C-reactive protein (CRP) was raised at 17.24 mg/L and erythrocyte sedimentation rate (ESR) was raised at 48 mm/hour. Biochemical screening was normal. A diagnosis of cellulitis was made. Patient was started on intravenous cloxacillin. Despite negative blood and swab cultures, the skin lesions showed no signs of improvement after five days of antibiotic.

Thus, skin biopsy was performed to rule out differentials such as intense contact dermatitis, intense insect bite reactions,

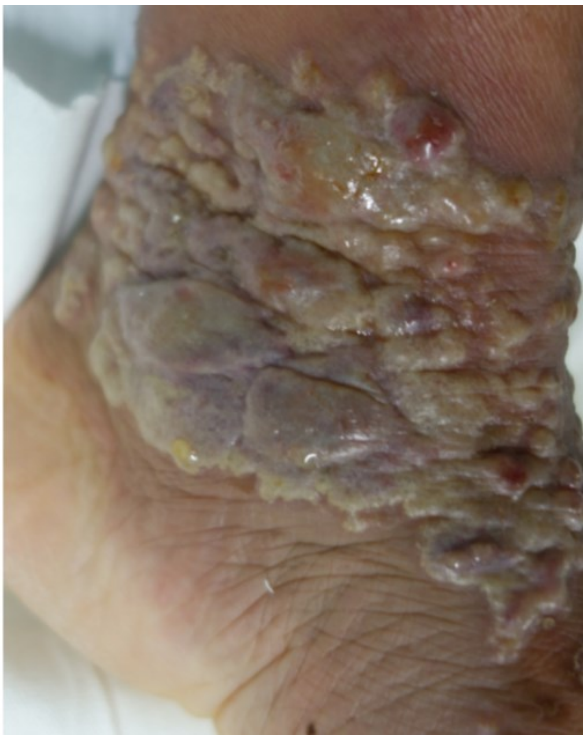

Figure 1: Tense blisters filled with serosanguineous fluid on the dorsal aspect of left foot.

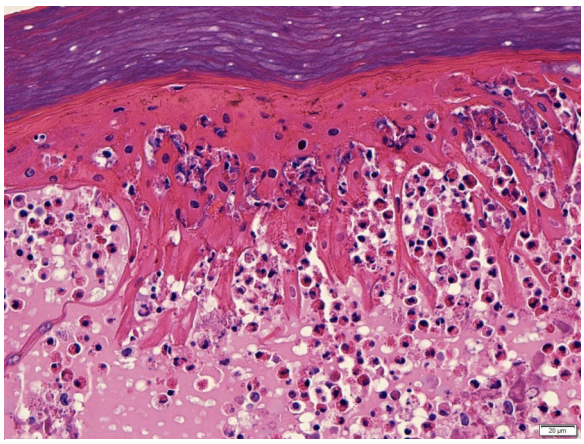

Figure 2: The epidermis showed marked spongiotic dermatitis with intraepidermal vesicles containing eosinophils mixed with occasional neutrophils (H&E stain, magnification x40).

bullous pemphigoid and eosinophilic cellulitis. Histological findings showed marked spongiotic dermatitis with intraepidermal vesicles containing eosinophils mixed with occasional neutrophils (Figure 2). The dermis showed a dense eosinophilic infiltrate mixed with scattered lymphocytes extending into the superficial part of the subcutaneous fat accompanied by foreign body granulomas (Figure 3a). In areas, flame figures were identified (Figure 3b). No subepidermal blister or features of vasculitis. No immune deposit was seen by direct immunofluorescence study. A diagnosis of bullous eosinophilic cellulitis was made.

Oral prednisolone 0.5mg/kg/day was commenced. After 72 hours, patient became afebrile with improvement of skin lesion and normalization of CRP and ESR. Prednisolone was tapered down over a course of two months, which resulted in complete resolution of skin lesions. Subsequent six-monthly follow up revealed no recurrences.

DISCUSSION

Bullous eosinophilic cellulitis or bullous Wells syndrome is a rare inflammatory skin disorder.[1, 2] Other variants of eosinophilic cellulitis include, plaque type, annular granuloma-like, urticaria-like, papulovesicular, papulonodular

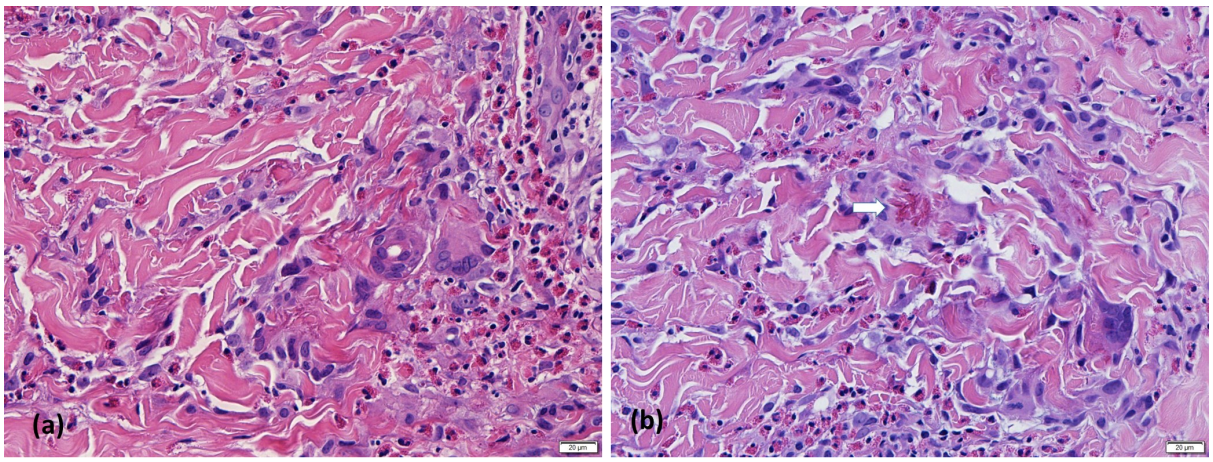

Figure 3: (a) The dermis showed a dense eosinophilic infiltrate mixed with scattered lymphocytes, accompanied by epithelioid cells granuloma, (b) Dense eosinophilic infiltrate with formation of flame figures (white arrow), (H&E stain, magnification x40).

and fixed-drug-eruption-like.[1]

Bullous Wells syndrome typically presents as an edematous and erythematous plaque, with presence of bullae.[2] The bullae can be tense or flaccid. It usually affects the extremities. The lesion can be painful and pruritic. Systemically, patient may complain of general malaise and fever. Clinically, it can mimic bullous cellulitis, intense arthropod bite, bullous pemphigoid or intense contact dermatitis.[7] Our patient presented with tense blisters filled with serosanguineous fluid and fever. Hence, a diagnosis of cellulitis was entertained initially.

The diagnosis of Wells syndrome is based on clinical history, examination, blood investigation and histological findings. Peripheral blood may reveal eosinophilia in approximately 50% of cases.[1] Besides supporting the diagnosis, eosinophilia may also help to indicate disease activity.[8]

Skin biopsy is crucial to diagnose Wells syndrome. Histologically, the biopsy shows a diffuse dermal infiltrate of eosinophils. The blisters can be intraepidermal or subepidermal. The most characteristic findings are flame figures, which are bright eosinophilic areas of collagen encrusted with eosinophil granules.[1, 2] The flame figures are sometimes surrounded by histiocytes and multinucleated giant cells. Although the presence of flame figure strongly supports diagnosis of Wells syndrome, it is not pathognomonic. It can only be detected in about 50% of cases.[1] Flame figure can be seen in other dermatoses with eosinophils, such as bullous pemphigoid, bites, drug eruption and eczema. But in the latter differential diagnoses discussed, epithelioid cell granulomas are not present together with the flame figures.

Bullous Wells syndrome has been reported to be associated with more sinister systemic conditions such as ulcerative colitis , lymphoma and Churg-Strauss syndrome.[3-6] Treating the underlying disease may lead to recovery of Wells syndrome.[3-5] Thorough history and examination involving respiratory and gastrointestinal systems, are mandatory in order to rule out systemic involvement. Our patient denied symptoms suggestive of systemic disease, hence further investigation was not warranted.

To date, there is no standard guideline to treat Wells syndrome. Although it can resolve spontaneously, Wells syndrome typically follows a relapsing and remitting pattern.[1, 2, 6] Steroid, both topical and systemic

are considered first-line treatment. Other therapies include dapsone and tacrolimus.[1, 2, 9] The prognosis is good, with resolution of lesion and no permanent scars. Our patient responded well to systemic steroid with no recurrences over one-year follow up.

CONCLUSION

Bullous Wells syndrome is a rare inflammatory skin disease. It can be misdiagnosed as other disease such as bullous cellulitis, which may result in delay in treatment. A comprehensive history, examination, blood investigation and histological review are mandatory to diagnose Wells syndrome. This case highlights the importance to suspect Wells syndrome in a patient with presumed cellulitis and eosinophilia who fails to respond to antibiotics. Misdiagnosis as other diseases may delay treatment and prolonged the course of the condition with increased morbidity.

CONFLICT OF INTEREST DISCLOSURES

There is no conflict of interest to be disclosed by all authors.

CONSENT

We have acquired consent for the above images to be used for publication purpose.

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