

CASE REPORT

An orange peel-like nodule on the back: A case of Wells syndrome

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Abstract

Wells syndrome is a rare eosinophilic syndrome, associating inflammatory lesions, suggestive histological images and frequent eosinophilia. Wells syndrome is characterized by multiplicity of anatomoclinical forms. Clinically, lesions may be urticarial, annular, papulonodular, papulovesicular or bullous. On histopathology, the flame aspect is by no means specific and late.

KEYWORDS

eosinophilia, eosinophilic syndrome, histopathology, Wells syndrome

1 | INTRODUCTION

Wells syndrome (WS) is a rare eosinophilic syndrome, associating inflammatory lesions, suggestive histological images, and frequent eosinophilia.¹ It is characterized by great clinical variability, which can delay diagnosis.² The typical presentation is the classic plaque-type variant that is often misdiagnosed as infectious cellulitis preferentially located on the extremities.³ Polymorphic cutaneous patterns were described: urticarial, annular, papulonodular, papulovesicular, and bullous.⁴ We report a case of WS with atypical presentation as an infiltrated orange peel-like nodule.

2 | CASE REPORT

A 39-year-old man presented with a one-week history of a mildly painful erythematous nodule on the lower

back. Examination revealed an infiltrated well-defined 3-cm nodule, with an “orange peel”-like appearance (Figure 1A). Millimetric satellite papules were noted (Figure 1B). Darier's sign was negative. Asthenia and headache had preceded the skin lesions' onset. Fever and inguinal lymphadenopathy had then appeared. There was no drug intake or insect bite. Complete blood count revealed eosinophilia, with a peak of $5.6 \times 10^9/L$, and C-reactive protein level was high (89 mg/L). Histopathological examination showed an eosinophilic infiltrate of the mid and deep dermis with few lymphohistiocytes and eosinophilic deposits. There was no vasculitis (Figure 2A, B). Investigations searching for a systemic involvement or an underlying etiology (cardiac ultrasound, viral serologies, thoraco-abdomino-pelvic CT scan, and stool examination) had not shown abnormalities. Based on clinical and histopathological features, the diagnosis of Wells syndrome was made. Outcome after doxycycline 100 mg daily and cetirizine 10 mg daily was unfavorable. The course was

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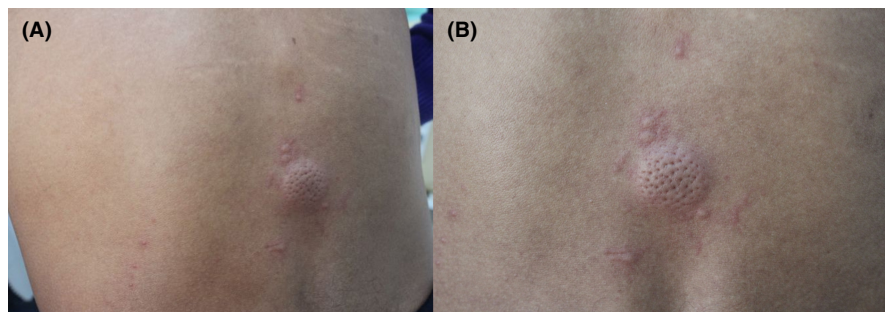


FIGURE 1 (A): Clinical features: A well-defined 3-cm nodule with an “orange peel”-like appearance (B) satellite papules.

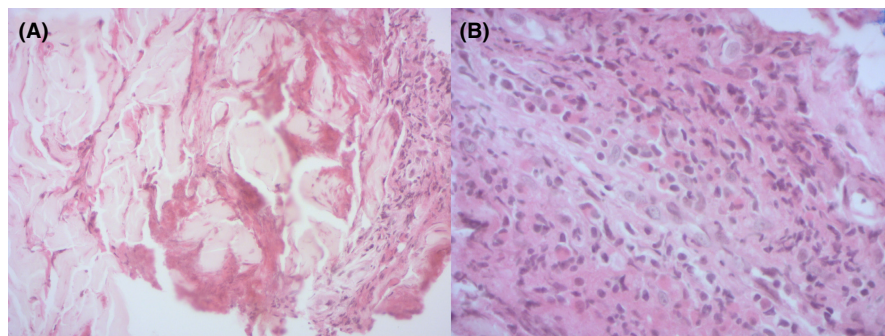


FIGURE 2 Histologic findings showing a deep dermal infiltrate composed of eosinophils and histiocytes around amorphous depositions of collagen (hematoxylin–eosin; (A): original magnification 200; (B): 400).

marked by a rapid clinical and biological regression after 2 weeks of prednisolone at a dose of 0.5 mg/kg daily.

3 | DISCUSSION

Clinical variability in Wells syndrome (WS) can delay diagnosis.² Polymorphic cutaneous patterns were described: urticarial, annular, papulonodular, papulovesicular, and bullous.⁴ The diversity of clinical aspects of Wells syndrome is probably explained by the level of eosinophilic infiltrate, which may be superficial or deep. To our knowledge, an orange peel-like nodule appearance indicative of the deep infiltration of inflammation has never been described in the literature. Systemic signs are rarely described. Histological flame-figure aspect, absent in our case, is not specific and had a delayed onset. The absence of vasculitis is an important negative sign. Many triggering factors have been reported: insect bites, infections, drugs, and underlying hematologic disorders.⁵ WS may be prior, revealing, or concomitant to these diseases. Long-term monitoring is essential. We report an idiopathic WS of misleading clinical presentation. We confirm the wide polymorphism of clinical and histological features of WS. This diversity seems to depend on the location of the dermal infiltrate.⁶ A variety of treatment options are described. The most common and effective treatments are oral steroids and dapsone.⁷ Other treatments such as doxycycline and colchicines are described such effective in some cases, but, due to the good

prognosis and tendency to resolve, systemic treatment should be limited to cases resistant to local therapy, especially with dermocorticoids.⁷

AUTHOR CONTRIBUTIONS

BM and SK wrote the paper. TM and ST wrote parts of the manuscript related to the histopathological aspects of the disease. MA contributed to the management of the patients and revised the article. HT critically reviewed the manuscript and gave final approval. All authors have read and approved the final manuscript and agree to take full responsibility for the integrity and accuracy of the work.

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None.

CONFLICT OF INTEREST

The authors have no potential conflicts of interest to disclose.

DATA AVAILABILITY STATEMENT

The data that support the findings of this study are available from the corresponding author upon reasonable request.

CONSENT

Written informed consent was obtained from the patient to publish this report in accordance with the journal's patient consent policy.

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