Eosinophil-Rich Sweet syndrome: Is it a new entity?

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Abstract

Sweet syndrome (SS) is an inflammatory disease with clinical and histological characteristics. SS is usually associated with important neutrophilic infiltrates on histological examination of skin biopsies. A few cases of SS with dense eosinophil infiltrate have been described. Herein, we report a new case of eosinophil-rich SS.

Introduction

Sweet syndrome (SS) is an inflammatory disease usually associated with an important neutrophilic infiltrate on histological examination of skin biopsies [1] A few cases of SS with dense eosinophil infiltrate have been described. Herein, we report a new case of eosinophil-rich SS in a woman with a history of chronic lymphocytic leukaemia (CLL).

Case report

A 65-year-old woman, diagnosed a CLL stage A of Binet, was stable for 5 years and has not require any specific treatment. She presented with febrile multiple erythematous and painful cutaneous lesions, three days prior to presentation. These lesions initially appeared on the buttocks, a few hours after an insect bite, with progressive extension to the skin sites. The patient had not received any new medications prior to the rash. Dermatological examination revealed multiple purplish erythematous plaques with different sizes varying from 1 to 3 cm, infiltrated borders, and centrifugal extension on the trunk, limbs, neck, and face (Fig. 1A and B). The palms of the hands were also affected (Fig. 1C). We did not observe any modifications in the surrounding area. The rest of the physical examinations revealed no abnormalities. Biological examinations were within normal levels despite the biological inflammatory syndrome. Histological examination of the cutaneous biopsy specimen revealed oedematous dermis associated with abundant perivascular and interstitial inflammatory infiltrates (Fig. 2A). This infiltrate was mainly composed of neutrophils associated with numerous polynuclear eosinophilic cells (PNEs) (Fig. 2B). She was treated with oral doxycycline at 100 mg/j in association with high-level topical corticosteroids (one application per day) for 10 days, resulting in clinical improvement and apyrexia from the 3rd day of treatment. The assessment of her haematological malignancy revealed a stable condition at stage A of Binet.

Discussion

Based on clinical, biological, and histological findings, a diagnosis of SS was established [1]; however, we found an inflammatory infiltrate rich in PNE. Indeed, the presence of some PNEs in association with neutrophilic infiltrates during SS has been well documented in the literature [2]. In a study of 73 cases of SS, the presence of PNE in association with neutrophilic infiltration was reported in 41% of cases [2]. However, eosinophil-rich SS is an exception. To the best of our knowledge, only three cases of eosinophils-rich SS have been reported [3, 4]. Among these three observations, one case was described in association with digestive T-cell lymphoma [4]. The clinical and histological features of the patients are summarized in Table 1.

In our case, the insect bite triggered a cutaneous eruption. So, the diagnosis of "exaggerated reactions to insect bites" also called "Eosinophilic Dermatoses of Hematologic Malignancies" (EDHM) can be evoked [5].

The present dermatosis seems to be an immunological reaction caused by sensitization to proteins in insect saliva [5]. It is characterized by one or multiple erythematous lesions, 2-10 cm in size, which appear a few hours after the insect bite with progressive extension and spontaneous healing after 3 to 10 days, without a febrile context [5]. Histologically, EDHM is characterized by the presence of numerous PNEs within the dermal infiltrate [5]. We suggest an overlap between paraneoplastic eosinophil-rich SS and EDHM. Therefore, we believe, that EDHM can be considered an eosinophil-rich SS.

Conclusion: We report an extremely rare case of SS characterized by rich dermal infiltration of eosinophils in association with CLL. Through our case report and the literature, we highlight the relationship between EDHM and paraneoplastic eosinophil-rich SS to make the clinician and the anatomopathologist aware of this new entity.

References

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Case, Author and Year of publication	Age (years)	Sex	Medical history/ Associated disease	Clinical presentation	Histology	Evolution	Triggering factor
N°1, Masuda et al, 1994	29	F	Asthma (prior to the diagnosis of SS)	Pruritic papules and plaques on face, neck, trunk and extremities Temperature: 37.7°C	Dense dermal infiltrate With multiple neutrophils and eosinophils.	Good evolution after the administration of 50 mg/day of dapsone. Recurrence of lesions 2 years after the first episod Prolonged treatment with Prednisolone (10 mg/day) and dapsone (100 mg/day)	Respiratory tract infection

Case, Author and Year of publication	Age (years)	Sex	Medical history/ Associated disease	Clinical presentation	Histology	Evolution	Triggering factor
N°2, Masuda, et al, 1994	61	M	None	Red papules and plaques necrotic center on face, neck and back. Fever : 39.3°C	Half of the cell infiltrate composed of neutrophils, eosinophils 10%.	Complete healing after 2 weeks of treatment with potassium iodide (l g/day) Recurrence at 2 months after the therapy, resolving under the same treatment for 7 days	None
N°3, Soon et al, 2016	90	M	Severe refractory celiac disease (Prior to the SS) Enteropathy-associated T-cell lymphoma (With the onset of SS)	Diffuse pink, oedematous, papules, nodules, and plaques on the back, buttocks and abdomen. Fever: 38.2°C	Superficial and mid-dermal perivascular and interstitial infiltrate with numerous eosinophils	Spread of the eruption Recurrent gastro-intestinal bleeding leading to death	None
Our case	65	F	Chronic lymphocytic leukaemia (2 years before the onset of SS)	Multiple erythema-tous and painful cutaneous lesions Fever (38.5°C)	Inflammatory infiltrate rich in eosinophilic polynuclear cells	Good evolution under doxycyline 100mg/day and high-level-topical corticos-teroid for 10 days (no recurrence with a follow-up for 2 years)	Insecte bite

Table 1 : Clinical and histological features of the 3 reported cases of eosinophil-rich Sweet Syndrome in comparison with our patient.

M : Male, F : Female

Legend of figures

Figure 1: A: Purplish erythematous plaques with infiltrated borders and centrifugal extension on the buttocks and the thighs. B: Purplish erythematous plaques with infiltrated borders on the face with peri-ocular involvement. C: Erythemato-oedematous plaques with infiltrated borders on the palms.

Figure 2. A : Oedema and inflammatory infiltrate of the dermis (Haematoxylin and Eosin X 100). B : Inflammatory infiltrate rich in eosinophilic polynucluar cells (Haematoxylin eosin X 400)