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ERYTHEMA ELEVATUM DIUTINUM AS MOST PROBABLE DIAGNOSIS: A CASE REPORT

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Abstract
Cutaneous vasculitis can be cause by multiple disorders or can be idiopathic. Many diseases can present with similar findings, therefore histopathologic examination is always require for confirming the right diagnosis. The erythema elevatum diutinum (EED) is a localized vasculitis, classified as a neutrophilic dermatosis. It's a rare cutaneous condition, distribute on the extensor surface of the extremities, more frequently in the dorsum of the hands, knees and elbows. They have a symmetric distribution and can be asymptomatic, painful; or pruritic, sometimes accompanied paresthesias. The most common clinical presentation is round erythematous papules which become erythemato-violaceous or purpuric plaques. There are not pathognomonic histopathological findings, but can present as a leukocytoclastic vasculitis with perivascular neutrophilic infiltration in the middle and superficial dermis. I presented the case of a 61-year-old female, with erythematous purpuric painful plaques, irregular, symmetric and elevated, located in both thenar regions of her hands and paresthesias. The patient's presentation is consistent with multiple characteristics of EED such as the description of the lesions, the anatomical location, the symmetric distribution and the histopathological findings of an initial disease. This patient does not have all the clinical progression and outcome, due to the initial stage of the disease.

Key words: cutaneous; erythema elevatum diutinum; leukocytoclastic vasculitis

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Introduction

Erythema elevatum diutinum (EED) is a rare disease. It's a vasculitis that affects the small vessels. The most common presentation is red-brown or violaceous papules and nodules over the dorsal hands. The disease is limited to the skin [1]. I am reporting an initial presentation of a case that the most probably diagnosis until now is the EED.

Case Report

A 61-year-old female, Hispanic, complaining of purpuric painful lesions in both of her hands, which develop in one day long. She report paresthesias in both hands and finger tips few days before the onset. She denies any other associated symptom as fever sensation, headache, chest pain, dyspnea, abdominal pain, vomiting, diarrhea, muscle ache or any other. But the last month she was very stress and possibly had some depression symptoms, for which she take clonazepam.

The clinical examination showed (Fig. 1, 2) erythematous purpuric plaques, which were irregular, symmetric, elevated lesions located in both thenar regions of her hands. Also between the main plaques were some blister-like lesions. In the periphery of the main plaque there were some satellite papules of similar color and consistency of the main lesion. There wasn't any other physical finding during the physical examination, including head, eyes, ears, nose, throat, neck, cardiovascular, chest, abdominal, lower extremities and neurologic examination.

The vital signs and laboratory workup was done: including CBC with platelets, coagulation studies, glucose, uric acid, VDRL, HIV ELISA, and urinalysis. There was every test within normal limits.

The preliminary diagnosis was vasculitis vrs pioderma. The patient was treated initially with IV clindamycin and topical aluminum acetate, with mild clinical response to treatment. Then she was treated with oral clindamycin, prednisone and Dapsone, causing improvement of the lesions within 3 days of treatment.

A smear, a gram stain and culture of fluid from the only pustule present was done and was negative for bacteria. The biopsy was done and showed: a dense interstitial neutrophilic infiltrate in the reticular dermis associated with large amount of nuclear dust. There are significant edema of the papillary dermis and some erythrocyte extravasation. Despite the dense infiltrate, there isn't fibrinoid necrosis in the vessel's wall. The epidermis shows hyper and parakeratosis, and the presence of a pustule. The changes describe may correspond to early changes of erythema elevatum diutinum although no frank vasculitis is observed, which would make the definitive diagnosis.

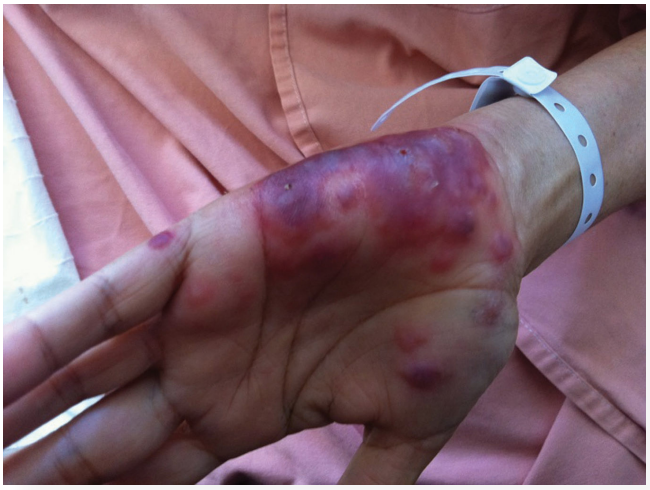

Figure 1. Left hand of the patient, showing the erythematous-purpuric plaque

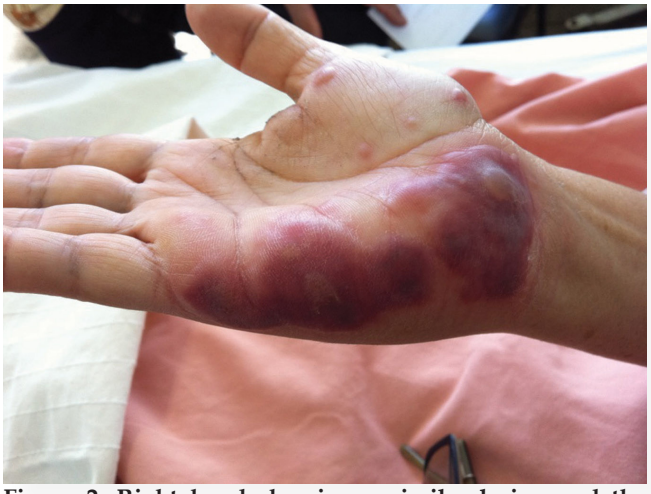

Figure 2. Right hand showing a similar lesion and the presence of the only pustule

Discussion

Cutaneous vasculitis can be cause by multiple disorders or can be idiopathic. Many diseases can present with similar findings, therefore histopathologic examination is always require for confirming the right diagnosis [1]. The nomenclature and classification are based on the histopathological findings. The etiology is still unknow [2]. The EED is a localized vasculitis and is classified into the neutrophilic dermatosis, due to the histopathologic findings [3].

Patients can present with multiple symptoms or physical findings, or can present only with the cutaneous manifestation. Accordingly you should perform multiple laboratory tests or radiologic studies based on the clinical findings of each patient [1].

The erythema elevatum diutinum (EED) is a rare cutaneous condition first describe considered a variant of leukocytoclastic vasculitis. The EED was first described by Hutchinson [4] in 1888 and Bury [5] in 1889. However the introduction of the name was proposed by Radcliffe-Crocker et al [6]. The name describes the lesions, erythema for redness, elevatum for elevated and diutinum for persistent or chronic [7]. The anatomical distribution is usually the extensor surface of the extremities, more frequently in the dorsum of the hands, knees and elbows. They have a symmetric distribution and can be asymptomatic, painful; or pruritic, sometimes accompanied by arthralgias or paresthesias [8].

The most common clinical presentation is round erythematous papules which become erythemato-violaceous or purpuric plaques. They can be accompanied of necrosis, vesicles and blisters. The clinical evolution of the disease is that the lesions can resolve spontaneously, resolve with treatment or can persist as chronic mild lesions with periodic exacerbations. Sometimes they can leave squeals as atrophy or depigmented areas. In addition with time the lesions can become hard nodules [9].

There are not pathognomonic histopathological findings, but can present as a leukocytoclastic vasculitis with perivascular neutrophilic infiltration in the middle and superficial dermis. The vessels can have a fibrinoid degeneration. The epidermis can be intact. With the time the biopsy can show the concentric perivascular fibrosis cause by the deposition of immune complexes. The old lesions have findings consistent with dermis fibrosis with multinucleated giant cells [9-14].

Although some patients have spontaneous resolution of symptoms, most of patients become chronic. The first treatment election is Dapsone which produces a suppressor effect on the lesions, is not curative and some patients have recurrence of symptoms when withdrawal. Some experts recommend 100mg per day as initial dose and then subsequent reduction until find the minimal ideal dose [15-17]. The Dapsone has severe secondary effects as metahemoglobinemia, hemolysis and agranolocytosis, which can be minimized with the combination of vitamin E or cimetidine in the beginning of the treatment. For resistant cases can use colchicine, tetracycline, niacinamida or nicotinamida. For the last option can use intralesional injections of corticoids [18,19].

Conclusion

The patient's presentation is consistent with multiple characteristics describe above as: the description of the lesions, the anatomical location, the symmetric distribution, the isolated cutaneous affection, the histopathological findings of an initial disease and the response to Dapsone. Obviously, the patient does not have the same clinical progression mention above, due to the initial stage of the disease, but I have to wait and see the clinical outcome.

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