CASE REPORT

Subcorneal Pustular Dermatosis (Sneddon Wilkinson Disease) in a patient with Rheumatoid Arthritis

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ABSTRACT

Subcorneal pustular dermatosis or Sneddon Wilkinson's disease is a rare entity, with unknown prevalence and incidence because it is underdiagnosed. It is characterized by soft sterile pustules with preferential location on the trunk and flexural regions such as the armpits and groin. The lesions can sit on healthy skin but are often found within a red patch. The pustules resolve in a few days and are replaced by fine scales before there is another relapse and new pustules form. Lesions may be pruritic, but are generally asymptomatic. It can get worse for a few weeks and then go away for months or years before reappearing. The process can be repeated for many years. The diagnosis consists of pathological, clinical and histopathological antecedents. To date, there are no defined criteria for the diagnosis of this disease, but it includes a variety of differential diagnoses that must be ruled out with an exhaustive study at the time of the anamnesis and the help of complementary tests.

We present the case of a 43-year-old male with a history of rheumatoid arthritis with dermatosis in flexural regions characterized by pustules, which was initially diagnosed as pustular psoriasis. However, considering the symptoms, histopathology and evolution, it was concluded that it was a pustular dermatosis subcornea or Sneddon Wilkinson's disease.

INTRODUCTION

Sneddon Wilkinson's disease is a rare neutrophilic dermatosis. Its pathogenesis remains unclear. So far, this condition's prevalence and incidence remains unknown. Mostly, it manifests in females in the fourth to seventh decade of life. It is associated to a great variety of comorbidities, such as autoimmune diseases and neoplasms. The etiopathogenesis identifies TNF-α as the main cytokine involved. Clinically, there is evidence of sterile pustules, characterized by the presence of "half-half" content in skin folds which can become generalized. The present diagnosis requires a holistic and meticulous approach. First line treatment corresponds to dapsone. Corticosteroids have brought positive results in some case reports.

A male patient presents with subcorneal pustular dermatosis in the basis of medical records, clinical and pathologic examination, along with complementary exams. A brief review of this entity is performed.

CLINICAL CASE

43-year-old male patient, diagnosed with rheumatoid arthritis less than a year ago, receives hydroxychloroquine and diclofenac treatment. The patient refers to the manifestation of skin lesions on axillary, inguinal folds and the elbow pit, associated with mild pruritus. Over time, lesions become more generalized and manifest on the anterior portion of the thorax and abdomen.

Physical examination reveals multiple pustules on purple erythematous patches located mainly on inguinal, axillary skin folds and the elbow pit (figure 1), as well as some disseminated pustules on the abdomen (figure 2). Half and half blisters are observed in greater detail (figure 3).

Routine laboratory exams show normal values and pustules without bacterial culture. The primary diagnosis, pustular psoriasis with psoriatic arthritis, required an HLA-B27 test, which turned out to be negative. Histopathologic examination evidenced a cleft between the corneal layer and granulosum layer, along with multiple neutrophil accumulation, scarce spongiosis and acanthosis, resulting in the following diagnosis: subcorneal pustular dermatosis. (figures 4 and 5).

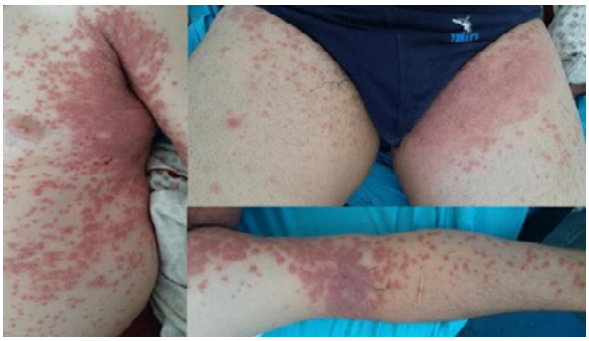

Figure 1: Pustular dermatosis on purple-erythematous base located on inguinal, axillary skin folds and the elbow pit.

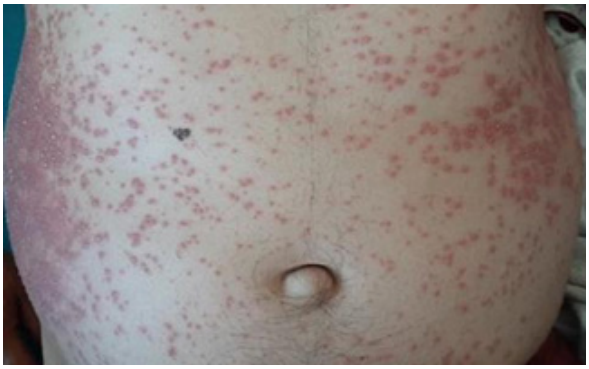

Figure 2: Disseminated pustules on the abdomen.

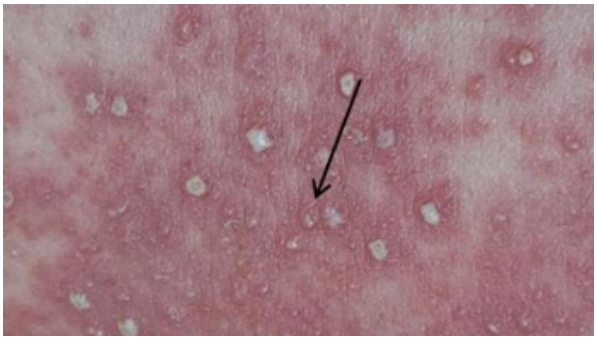

Figure 3: Half and half blister.

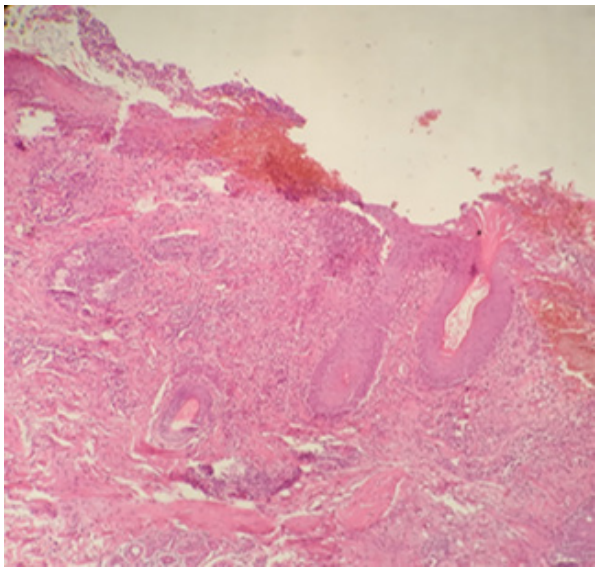

Figure 4: Subcorneal intraepidermal cleft, no spongiosis and scarce acanthosis (H/E 10x).

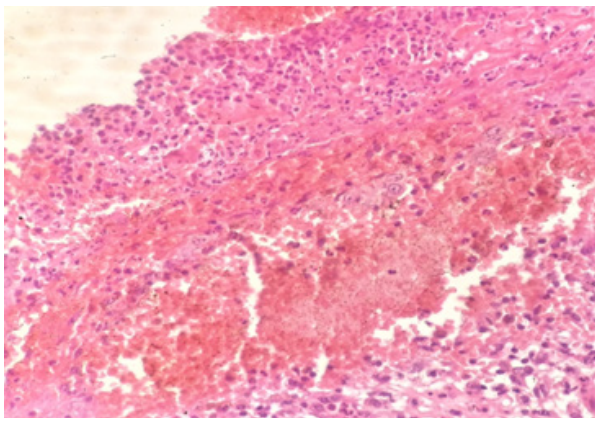

Figure 5: Close-up reveals neutrophil infiltration (H/E 20x).

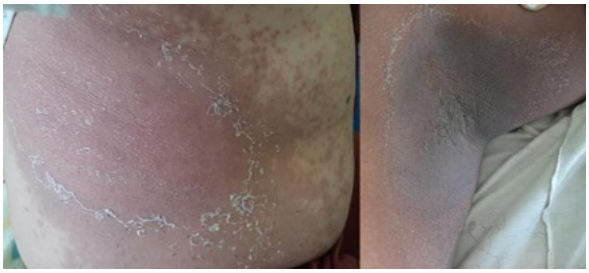

Figure 6: Postinflammatory pigmented, desquamative lesion.

The treatment during admission corresponds to 40 mg/day of oral corticosteroids. At the 15-day-follow-up, the patient shows favorable results: resolution of pustules and erythematodesquamative macules with discrete postinflammatory pigmentation (figure 6).

DISCUSSION

Subcorneal pustular dermatosis, also known as Sneddon and Wilkinson disease, is a neutrophilic dermatosis firstly described in 1956 by Sneddon and Darrel Wilkinson themselves. It is a recurrent and benign pathology, associated with a variable spectrum of pathologies: immunologic, neoplastic, inflammatory, among others.[1]

Epidemiologically, it is a rare and underdiagnosed disease; reason why its prevalence and incidence is unknown.[1]

It commonly manifests between the fourth and seventh decade of life, mainly on female adults. However, there are some cases involving pediatric patients.[2]

Being a neutrophilic disease, its etiopathogenesis is bound to an exaggerated activation and migration of neutrophils in the subcorneal layer, followed by an increase of proinflammatory cytokines, such as the IL6, IL8, IL10, C5a activation, mediated by the IgA and TNF-α liberation. Subcorneal pustular dermatosis is associated with a great variety of immunologic diseases, whose action mechanism is also bound to TNF-α activity, such as rheumatoid arthritis, inflammatory bowel disease, monoclonal gammopathies, and multiple myeloma.[1,2,3]

Clinically, this condition is characterized by sterile pustules located on axillary, inguinal skin folds, and on the neck and lower abdomen. Mostly, it manifests symmetrically, without affecting acral areas like the feet or hands. It does not affect mucosal areas either. Pruritus is present, though not in an excessive manner. The essential feature of pustules is the sign described as “half-half” or “hypopyon pustule”, where there is a fluid-filled serous superior portion and a deep or dense lower portion of purulent material. These pustules may aggregate.[1,4,5]

Histopathology indicates subcorneal neutrophil accumulation without the presence of microorganisms. There is little to no presence of spongiosis and acanthosis. Early stages of the condition evidence perivascular neutrophil migration across the epidermis. Histopathologic alterations are not specific. Consequently, an integral clinical study of the patient is required.[2,3,6]

Among the main differential diagnoses are the IgA pemphigus, of which the immunofluorescence revealed small deposits. Although some authors identify the entity as a pemphigus, purists rely on the immunofluorescence to make a differentiation. Differential diagnosis with pustular psoriasis is made through histopathology, which evidences spongiosis and marked acanthosis; this type of psoriasis may be related to psoriatic arthritis. In consequence, we ruled out such diagnosis though an HLA-B27 study. Another entity is the dermatitis herpetiformis. This condition presents with tense vesicular lesions, commonly located on extension regions, such as wrists and elbows. It produces intense pruritus, which is not something characteristic of subcorneal pustulosis. Histopathology reveals neutrophilic and eosinophilic infiltrate in papillary dermis. Acute generalized aexanthematous pustulosis may also be considered. This entitiy also requires to look out for certain medications due to drug reaction.[3,4,8,9]

First line treatment includes dapsone (100mg/day). After 4 weeks, resolution is achieved. To prevent an upsurge, minimum maintenance doses are required. Other antineutrophilic drugs include colchicine, sulphapiridine and cotrimoxazol, due to its immunomodulation mechanism. The use of topical corticosteroids associated with dapsone or the exclusive use of systemic corticosteroids was successful according to some

case reports. Furthermore, according to other studies, the administration of oral retinoids, such as acitretin resulted in an 8 to 15 day resolution. Moreover, there is a report that indicates the use of phototherapy and dapsone or retinoids. Since it is a TNF-α mediated disease, biologic therapy, such as infliximab and etanercept, has proven to be effective.[1,3,8]

When diagnosing this entity, it is essential to consider associated comorbidities. A 30-year-old research by Watts shows that subcorneal pustulosis is associated to numerous systemic conditions, such as connective tissue diseases (seropositive and seronegative rheumatoid arthritis), systemic lupus erythematosus, Crohn's disease; and hematologic disorders such as, aplastic anemia, multiple myeloma; and neoplastic diseases such as lung epidermoid carcinoma and apudomas metastatic. In their research involving 10 patients, Aguilera et al report an association of subcorneal pustulosis with arthropathy; seven comply with the requisites of the American Rheumatism Association and three with seronegative arthropathy. Currently, regarding the etiopathogenesis, TNF-α is the most involved cytokine. Two patients experienced an increase in the levels of this molecule, both regarding the serum and the pustule content.[6,7,8]

Lastly, Ackerman acknowledged the entity as "sui generis," and stated the condition was an evolutionary phase of pustular psoriasis, while Calonje et al consider the Sneddon Wilkinson disease as a different entity from psoriasis. Bordignon highlights that since the condition is underdiagnosed and undefined, prevalence is not well known, that which limits the conduction of further studies.[6,7,9,10]

CONCLUSION

The objective of this presentation is to inform about a rare and undefined entity. There are some discrepancies about its classification as an independent entity or as part of pustular psoriasis. We are inclined towards the latter.

In spite of showing unknown prevalence and incidence rate, some cases of the condition are described in the literature as being associated with rheumatoid arthritis. Furthermore, we must not forget that arthropathy is not only related to subcorneal pustulosis, but also to pustular psoriasis. The most essential aspect lies in acknowledging concomitant diseases. When suspicions of the Sneddon Wilkinson disease arise, studies must be demanded at the right time, along with an appropriate multidisciplinary procedure.

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