

Subcorneal pustular dermatosis (Sneddon-Wilkinson disease) during a pregnancy: unusual associations

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Abstract

Sneddon-Wilkinson disease, is a chronic relapsing vesiculopustular eruption typically seen in female after the fourth decade. Cutaneous lesions are generally found on the trunk and in intertriginous areas, and pustules may coalesce to form circinate patterns (1). We present a case of Sneddon-Wilkinson's disease in a 36-year-old woman when she first presented in dermatology at 24 weeks of pregnancy. In this discussion, we explain the pathophysiology and clinical presentation of Sneddon-Wilkinson's disease. Various medical conditions that have been associated with Sneddon-Wilkinson's disease are discussed. We describe systemic and topical treatment options for the treatment of Sneddon-Wilkinson's disease, the first of which is systemic dapsone. This patient is an excellent case of Sneddon-Wilkinson's disease with an unusual association.

Keywords: sneddon-wilkinson, hypercalcemia, pregnancy

Introduction

Subcorneal pustular dermatosis (Sneddon-wilkinson disease) is a rare condition that belongs to the neutrophilic dermatosis family. It is characterized by a amicrobial pustular rash. Its association with hypercalcemia and pregnancy remains unusual.

Case report

We report the case of a 36-year-old patient pregnant with 24 weeks, with no pathological history, who reported a pustular rash 3 months before admission. The back and left breast, trunk, lower limb root and thighs were covered with flaccid and tense pustules of variable size, yellowish, non-follicular, resting on an erythematous base. These pustules were associated with slightly painful crustal erosions. Our patient was in good general condition and was afebrile.

In front of this clinical picture, several diagnostic hypotheses were evoked: impetigo herpetiformis, dermatitis herpetiformis, pustular psoriasis, Sneddon-Wilkinson's pustulosis. The result test of bacteriological sample was sterile. The histology objectified an undercorneal detachment, of pustular appearance, rich in neutrophils, associated with neutrophil exocytosis, leading to the diagnosis of Sneddon-Wilkinson undercorneal pustulose.

Hypercalcemia at 120mg/l was found during repeated biological examinations, revealing hyperparathyroidism, confirmed radiologically by the presence of 2 parathyroid nodules.

The patient received surgical treatment by removing adenomas. After delivery, 100 mg/day disulone was introduced with good clinical improvement and early healing of lesions.

Discussion

Subcorneal pustular dermatosis (SPD) is a rare, chronic, relapsing pustular eruption within the subcorneal layer of the skin (2). The exact etiology is unknown (2). It mostly affects middle-aged women. It is characterized by aseptic

flaccid nonfollicular pustules surrounded by a thin erythematous areola and grouped in a circular fashion. A fluid or hypopion level may be visible within the largest pustules. SPD predominates on the trunk and proximal extremities and favors intertriginous regions (3). It may coexist with systemic diseases such as monoclonal gammopathies, pyoderma gangrenosum, multiple myeloma, rheumatoid arthritis and systemic lupus erythematosus (3). Hypocalcemia is also usually found, unlike hypercalcemia, which should encourage the search for an underlying cause, as was the case with our patient. In addition, it appears that pregnancy may be a trigger factor for sneddon pustulose, as pregnancy is considered a pro-inflammatory condition by the increase in inflammatory cytokines, especially after 20 AS.



Fig 1: (a) The patient's trunk at the initial presentation and (b) the patient's back at the initial presentation.

Histologically, subcorneal pustular dermatosis is characterized by subcorneal neutrophilic abscesses. Rarely, acantholytic cells and eosinophils may be seen within the blister. A mixed superficial perivascular infiltrate is usually present. Most cases of subcorneal pustular dermatosis have not, however, revealed positive immunofluorescence staining patterns (4).

Disulone remains the treatment of choice and helps to control this chronic relapsing-remitting condition. In contrast, it has been established as an antimicrobial/antiprotozoal agent by inhibiting the synthesis of dihydrofolic acid and resembling the action of sulfonamides (5). It also acts as an anti-inflammatory agent due to its ability to scavenge reactive oxygen species and an immunomodulator by suppressing interleukin-8 and tumor necrosis factor- (5).

Conclusion

Sneddon-Wilkinson's subcorneal pustulosis remains a rare pathology, with chronic and recurrent progression, but with a benign prognosis. Our observation is original because of the unusual association of snedden subcorneal pustulosis with hypercalcemia and pregnancy.

References

1. Kasha EE Jr and Epinette WW. Subcorneal pustular dermatosis (Sneddon-Wilkinson disease) in association with a monoclonal IgA gammopathy: a report and review of the literature. *J Am Acad Dermatol.* 1988; 19(5 Pt 1):854-858.
2. Watts PJ, Khachemoune A. Subcorneal pustular dermatosis: a review of 30 years of progress. *Am J Clin Dermatol.* 2016; 17(6):653-671.
3. Reed J, Wilkinson J. Subcorneal pustular dermatosis. *Clin Dermatol.* 2000; 18(3):301-313.
4. Sneddon IB, Wilkinson DS. Subcorneal pustular dermatosis. *Br J Dermatol,* 1979; 100:61-8.
5. Wozel G, Blasum C. Dapsone in dermatology and beyond. *Arch Dermatol Res.* 2014; 306(2):103-124.