Subcorneal Pustular Dermatosis of Sneddon-Wilkinson

Subcorneal pustular dermatosis is a rare, chronic, relapsing pustular eruption that strikes the corneal layer of the skin and presents with a neutrophilic infiltrate on histopathological examination. It was first described by Sneddon and Wilkinson in 1956 and affects mainly middle-aged and elderly women. Its coalescing vesiculopustular eruptions typically involve the flexion sites of the trunk and proximal extremities.

Its etiology is not fully understood, presenting several pathophysiological mechanisms, one of them being increased tumor necrosis factor-α. It is frequently related to systemic pathologies, particularly hematological (gamopathies, multiple myeloma) and inflammatory diseases (rheumatoid arthritis).

In this report is described the clinical case of a 74-year-old woman with metabolic syndrome who was hospitalized due to an extensive rash involving trunk and upper limbs associated with constitutional symptoms for a month.

Dermatology collaboration was requested, and solitary by clinical observation and attending to the presence of classical signal which consists of superficial pustules arranged in annular and serpiginous pattern (see picture) classified the eruption as a Sneddon-Wilkinson dermatosis. This diagnosis, has indication to further study associated systemic pathologies and medical therapeutic intervention with dapsone and local wound disinfection.

Of the complementary diagnostic tests performed, an increased sedimentation rate of 30 mm and an IGA gammopathy, serum values of 1118 mg / dl confirmed in immunofixation and protein electrophoresis, and were considered as positive findings. With normal hematological formula and without other focusing clinic the patient was discharged from the hospitalization for follow-up in outpatient internal medicine and dermatology.

Despite a notorious improvement in investigation techniques, the pathogenesis of this dermatosis is still controversial. It was found that some patients with Sneddon and Wilkinson’s disease show epidermal intercellular immunoglobulin A (IgA) deposits on direct and indirect immunofluorescence, which places them in the group of IgA pemphigus.

The diagnosis is still established if the classic signal is present (see picture), or if there is confirmatory histology. Disease without cure, dermatological control’s treatment and prognosis is dependent on systemic associations.

BIBLIOGRAPHY
4. Hashimoto T, Teye K, Ishii N. Clinical and immunological studies of 49 cases of various types of intercellular IgA dermatosis and 13 cases of classical subcorneal pustular dermatosis examined at Kunume University. British Journal of Dermatology, June 2016, 3.