TWO FAMILIAR CASES OF ACNE CONGLOBATA-HIDRADENITIS SUPPURATIVE ASSOCIATED TO PYODERMA GANGRENOSUM

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INTRODUCTION:

The so called "follicular occlusion triad" (FOT) or "appocrine acne" includes as clinical manifestations: Acne conglobata, hidradenitis suppurativa and perifolliculitis abscedens et suffodiens.

This entity is sometimes associated to joint-bone diseases, the combination of both manifestations (skin and skeletal) originate syndromes such as SAPHO (spondyloarthropathy, acne, pustulosis palmare et plantaris, hyperostosis and osteitis) or PAPA (pyogenic arthritis, pyoderma gangrenosum and acne conglobata); seeming to have genetic transmission (not only based on MHC system but also in genes isolated from different chromosomes as 15q).

Manifestations from follicular occlusion triad like acne conglobata (AC) have been associated to pyoderma gangrenosum (PG): 2 out of 86 patients with PG developed AC (Powell et al.); and 3 out of 10 patients with AC also presented PG (Rosner et al.). And both FOT/PG sometimes are linked to articular manifestations.

In spite of these findings there is no clear genetic transmission described for FOT manifestations neither is for PG; except for hidradenitis suppurative or skin-skeleton syndromes already mentioned above, that seem to have genetic factors involved.

CASE REPORT:

We report two familiar cases (brother and sister):

1) Woman 39 years old with a history of acne during adolescence, developed nodule-cystic lesions occasionally suppurative in both axillary folds (Fig 1) and inguinal-pubic area (at the age of 32 years old). Based on clinical features was diagnose of hidradenitis suppurativa being treated with antiandrogenic therapy and antibiotic drugs with good evolution. At the age of 37 she presented a flare of purple-erythematous plaques partially ulcerated in the upper zone of both thighs (Fig 2), suggesting clinically pyoderma gangrenosum (that was confirmed with a biopsy), and improving with oral steroids.

2) Man 43 years old, he had been showing since puberty nodules-cystic lesions in the upper trunk, thorax and neck, that evolved to keloid scars (Fig 3, 4). In addition to this lesions appeared inflammatory-suppurative plaques in both axillary (Fig 5) and inguinal folds. All this suggested acne conglobata associated to hidradenitis suppurativa. In the back were observed round vegetating lesions resembling pyoderma vegetans (Fig 6).

DISCUSSION:

Here are reported two familiar cases with clinical manifestations of FOT (except for perifolliculitis capitis abscedens et suffodiens): not associated to date with articular manifestations, and at least in one of them associated to PG. No previous descriptions of several FOT manifestations associated to PG in members of the same family have been found. This could be explained as an incomplete variant or a non completely evolved skin-articular syndrom (although articular manifestations usually precede skin ones).

5) Shamsaf P.D., Toma M.S. Pyoderma gangrenosum associated with cystic acne and hidradenitis suppurativa controlled by adding minocycline and sulfasalazine to the treatment regimen. Cutis. 1996; 57: 315-319