

Pyoderma Gangrenosum with an Atypical Location in a Rare Diagnosis

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Sir,

A 27 year old patient without any notion of unprotected sexual intercourse or known inflammatory pathology, admitted for consultation for the management of a progressive genital ulcer for more than 3 months, painful, the clinical examination had objectified the presence of a ulcer of 4 cm taking the whole glans, surmounted by meliceric crusts in some places and hemorrhagic by others, foul-smelling with issue of pus at pressure, the lymph nodes were free, especially inguinal. This clinical aspect was more in favor of a pyoderma gangrenosum with genital location (**Figure 1**), the rest of the clinical examination had objectified the presence of several erythematous papulo-nodules on the level of the back, buttock, pubis and the trunk (**Figures 2 and 3**), as well as retentional lesions on the level of the face, associated with cysts on the level of the chin and the nape of the neck (**Figure 4**), by pushing the interrogation the patient reported the notion of swelling of the left ankle joint at the age of 12.

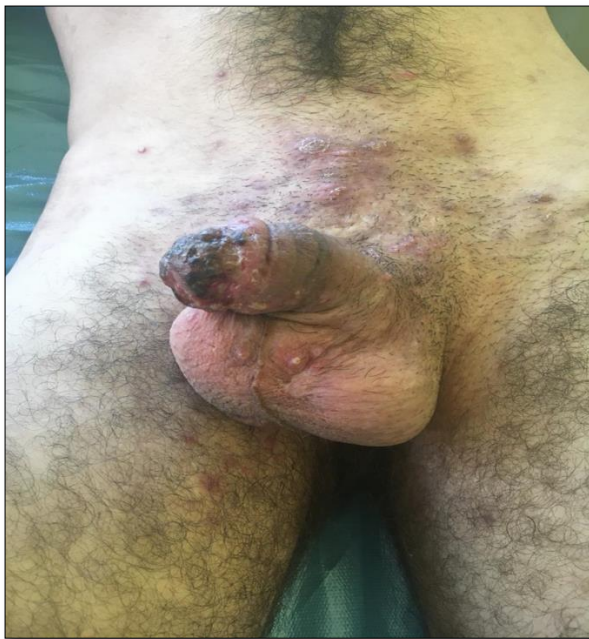


Figure 1. Pyoderma gangrenosum of the penis.

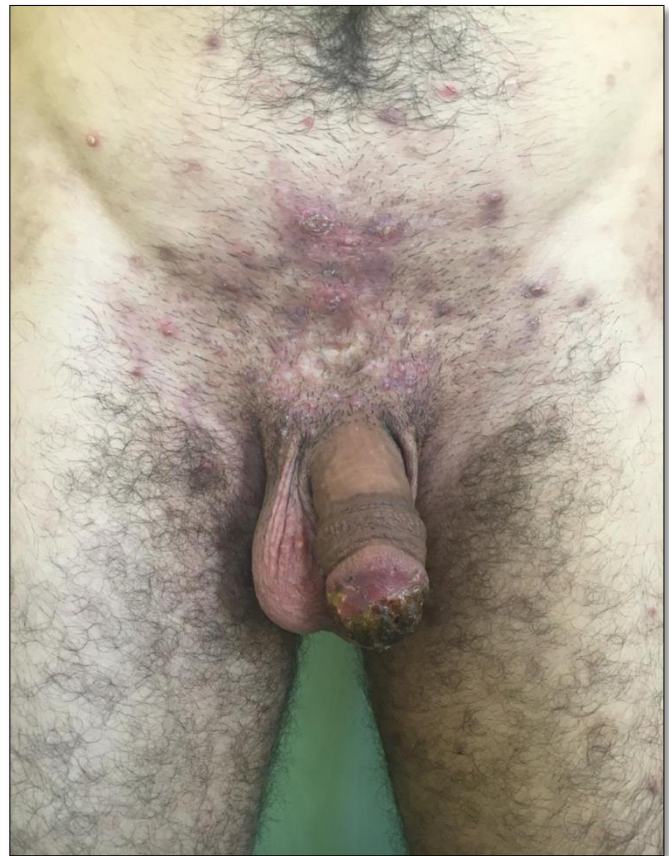


Figure 2. Pyoderma gangrenosum associated with acne nodules in the pubis.

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Figure 3. Papulopustular back acne.



Figure 4. Cysts and nodules in the neck.

Given this clinical presentation and the patient's history, the diagnosis of PAPA syndrome (pyogenic arthritis, pyoderma gangrenosum and acne) was made and the patient was put on a combination of systemic corticosteroids (CO) and isotretinoin low dose for 15 days then stop CO and gradual increase in retinoid doses with good progress, scarring of genital ulceration and improvement of acne lesions.

Pyoderma gangrenosum (PG) is a rare sterile inflammatory neutrophilic dermatosis [1] characterized by recurrent skin ulcers with mucopurulent or hemorrhagic exudate, which can be reactive to or associated with several pathologies [2].

PAPA syndrome is a rare inflammatory syndrome considered as an acne syndrome, grouping together arthritis often appeared in pre-pubertal age, pyoderma gangrenosum and severe nodulocystic or conglobata acne [3]; its treatment is sometimes difficult requiring high doses of retinoids and sometimes even use more severe therapies such as anti TNF alpha [4].

In our patient, genital ulceration was the sign which pushed him to consult, and the diagnosis could be diverted towards an infectious or even tumor cause given the chronicity of the evolution, and a good clinical examination with a precise interrogation allowed to correct the diagnosis, although, one of the criteria for the diagnosis of the PAPA syndrome which is arthritis was not objectified in our patient at our clinical examination and we just based on the history and the symptoms reported by the patient to describe this involvement, it must be said that it is currently difficult to say whether old arthritis can be included in the diagnosis of PAPA syndrome, but given the context and the association of genital pyoderma gangrenosum with nodular acne lesions, we have taken into consideration this notion of arthritis.

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