



PAEDIATRIC DERMATOLOGY

ASSOCIATION OF PYODERMA GANGRENOSUM, CROHN AND INCONTINENTIA PIGMENTI IN A CHILD

S Elloudi⁽¹⁾ - H Bay Bay⁽¹⁾ - S Zinoune⁽¹⁾ - Z Douhi⁽¹⁾ - Fz Mernissi⁽¹⁾

University Hospital Center, Dermatology, Fez, Morocco⁽¹⁾

Introduction: Pyoderma gangrenosum (PG) is a rare neutrophilic dermatosis of unknown cause, occurring readily in middle-aged women, characterized by locoregional aggressiveness and chronic, recurrent development.

Incontinentia pigmenti (IP) is a rare genodermatosis of X-linked dominant transmission, classified as neurocutaneous syndromes.

We report the case of a 7-year-old girl being followed for incontinentia pigmenti with severe perioral pyoderma gangrenosum associated with Crohn's disease.

Observation: A 7-year-old girl, born of a non-consanguineous marriage, followed since birth for incontinentia pigmenti, presenting since one year oral and perianal ulcerations evolving by thrust remission, rebels to the local treatment and antibiotics, consulted at our service for a rapidly extensive perioral ulceration that has been evolving for 20 days. In addition, the patient reported inflammatory gonalgia without other signs including digestive and neurological. Dermatologic examination revealed an ulcerated tumor of 5cm long axis interesting the lower lip, overlying the right peri-oral region, well limited irregularly-shaped by place, infiltrated base, and purulent surface covered with meliceric and haemorrhagic crusts. Oral opening was limited. Follicular and non-follicular pustules sitting on the legs, a macroscopic dyschromic patch with a cribriform appearance in the perianal, linear pigmented cicatricial stria along the lines of blaschko at the level of the trunk and limbs. The biological assessment revealed a syndrome and an inflammatory anemia. A skin biopsy of perioral ulceration showed a polymorphic inflammatory infiltrate with a predominance of PNNs consistent with PG. Digestive exploration with histological study was in favor of Crohn's disease. The patient was on steroids at 0.5 mg / kg / day with regression of ulceration.

Conclusion: The association of PG and crohn is classic, the antecedent of IP can make discuss a possible link between these three pathologies.

