INFLAMMATORY SKIN DISEASES (OTHER THAN ATOPIC DERMATITIS & PSORIASIS)

PYODERMA GANGRENOSUM ASSOCIATED WITH BECHET'S DISEASE: A SERIES OF 5 CASES

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Background: Pyoderma gangrenosum (PG) is an autoinflammatory disorder that presents as rapidly enlarging painful ulcers, which are often idiopathic or can be associated with underlying systemic disorders such as inflammatory bowel disease, various blood dyscrasias and PAPA syndrome amongst others. Less commonly, it has been described in people with Bechet's Disease (BD). BD is a rare multisystem inflammatory disease which has traditionally been associated with people who lived along the Silk route. It is characterised by a constellation of symptoms which often start with the occurrence of oral and genital ulceration, which usually precedes diagnosis. The treatment of both these entities is guided by clinical activity with immunosuppressant therapy. To date there are fewer than 5 case reports describing the association between PG and BD.

Observation: We describe five cases of PG occurring in patients with BD, where investigations for other underlying causes including inflammatory bowel disease, were negative. PG occurred after a diagnosis of BD in 4 of our cases and in the other was described alongside orogenital ulceration at time of diagnosis. In 2 cases, PG resolved or improved on successful immunosuppression of BD activity and in the remaining cases PG remained active despite treatment, which mirrored BD disease activity. All these patients also suffered from end-organ damage as a result of their BD.

Key Message: Currently PG is not thought to be associated with BD, however, our cases have demonstrated that there is very close clinical correlation between BD disease activity and severity of PG. This suggests that they may share a similar pathophysiology and although further studies are needed, PG maybe considered as part of BD in select cases.