P304

P304 -Relapse of ANCA-associated vasculitis with recurrence of pyoderma gangrenosum and de novo pulmonary haemorrhage

Dr Matthew Beresford[1], Dr Chris Duhovic[2], Professor Alan Salama[1], Dr Sally Hamour[1]
[1]Royal Free Hospital, London, United Kingdom, [2]Whittington Hospital, London, United Kingdom

Pyoderma gangrenosum (PG) is an unusual skin disorder which usually occurs either in isolation or associated with systemic disease such as inflammatory bowel disease or haematological malignancy. Whilst skin manifestations in patients with ANCA-associated vasculitis (AAV) are common, pyoderma gangrenosum is rarely seen in association with AAV. We report a case of relapse of PR3-AAV with new pulmonary haemorrhage, heralded by the deterioration of previously quiescent pyoderma gangrenosum.

A 69 year old woman presented with large, painful, enlarging ulcers on her lower limbs. After clinical assessment and skin biopsy, she was diagnosed with primary PG and commenced on azathioprine and high-dose oral steroids. Six months later she re-presented with a relapse of PG, associated with acute polyarthritis, haemoproteinuria and an ANCA-PR3 titre of >100. Renal biopsy confirmed a healing pauci-immune glomerulonephritis with fibrocellular crescent formation. She received induction treatment for AAV with two doses of rituximab and further high-dose steroids with an excellent clinical response, including the skin lesions.

Two years later, she presented acutely with haemoptysis and shortness of breath. In the two months prior, she described enlargement and ulceration of a previously quiescent left lower leg skin lesion. Examination of the skin revealed a 5cm by 10cm tender, violaceous skin lesion with central ulceration on the left lower leg. Laboratory testing revealed severe anaemia with an acute phase response and extensive air-space shadowing in the lungs on imaging. She was diagnosed with AAV-associated pulmonary haemorrhage, with associated relapse of PG, and was treated with steroids, cyclophosphamide and a course of plasma exchange. By day ten, the patient's dyspnoea and haemoptysis had resolved, her skin lesions had begun to improve and she was discharged home.

AAV is occasionally associated with PG and it is therefore important that ANCA-testing and clinical assessment for systemic vasculitis should be considered in these patients. Presentation or relapse of PG may significantly pre-date that of systemic disease. AAV has also been reported in association with other neutrophilic dermatoses such as Sweet's syndrome and mechanisms of neutrophil dysregulation might provide a clue to these associations.