

**VASCULITIS IN A PATIENT WITH ALPHA-1-ANTI-TRYPsin DEFICIENCY SUCCESSFULLY TREATED WITH CYCLOSPORINE AND TOPICAL NEGATIVE PRESSURE**

Kirsten Gabriel, Karsten Fogh

*Department of Dermatology, Aarhus University Hospital, Aarhus, Denmark*

**Aim:** Cutaneous vasculitis is a severe skin condition rarely associated with alpha-1-anti-trypsin deficiency. We demonstrate the beneficial effect of cyclosporine and topical negative pressure (TNP) on non-healing vasculitis in a patient with severe lung insufficiency related to alpha-1-antitrypsin deficiency.

**Methods:** A 54 year old woman with a 15 year history of lung insufficiency related to alpha-1-anti-trypsin deficiency developed multiple lesions of both tibiae in July 2007. Clinical examination and histology showed changes consistent with vasculitis. Topical corticosteroid treatment and compression therapy resulted in healing of a number of lesions except two vasculitic that increased in size and wound infection developed together with severe wound pain. Due to previous side effects of systemic corticosteroids, other treatment options were considered.

**Results:** Cyclosporine (3 mg/kg/day) was initiated November 2007 together with intravenous antibiotics and strong opiates. Improvement of inflammatory changes was observed but healing did not improve and the patient developed denuded tibia bone (2 cm<sup>2</sup>) in the wound bed on the right side. TNP was applied for 5 weeks resulting in marked healing of the two ulcers. Cyclosporine was stopped after 12 months. Healing occurred on the right side and in January 2008 a 2 cm<sup>2</sup> superficial ulcer was present on the left side.

**Discussion:** This case demonstrates beneficial effect of cyclosporine and topical negative pressure on healing of cutaneous vasculitis in a patient with severe pulmonary insufficiency caused by alpha-1-antitrypsin deficiency.