To our knowledge, this is the first report of a successful split thickness skin graft to a large surgical wound in a patient with scleroderma. In this case history, the key point is a clinical one—that wound healing may be uncomplicated even if the graft is taken from affected sclerodermatous skin and transplanted to another affected area. There are reports in the literature of using artificial [8], split thickness [9] and autologous [10] skin grafts to promote ulcer healing in patients with SSc.

In conclusion, in this case, primary closure of the wound was impossible, and there was a real concern that our patient might have required amputation. Grafted skin presented a natural barrier for preventing infection and allowed early wound healing. Our patient thus demonstrates how split thickness skin grafting even from sclerodermatous skin can be successfully used to cover large wounds with a good outcome in patients with SSc.

Disclosure statement: The authors have declared no conflicts of interest.

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Accepted 21 November 2007
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Rheumatology key message

- Autologous skin graft, even involving affected skin, can facilitate wound healing in systemic sclerosis.

Rheumatology 2008;47:380–381
doi:10.1093/rheumatology/kem330
Advance Access publication 31 January 2008

Crohn’s disease, relapsing polychondritis and epidermolysis bullosa acquisita: an immune-mediated inflammatory syndrome

Sir, We report the case of a 22-yr-old Caucasian woman admitted to hospital due to frank haemoptysis associated with cutaneous lesions in hands and feet, congestion of the upper airway tract, episcleritis and impaired phonation. She had a 2-yr history of Crohn’s disease (CD), diagnosed by colonoscopy and intestinal biopsy, in remission under methotrexate treatment (12.5 mg/week). She referred progressive association of bilateral eyelid retraction with epiphora, nasal congestion with saddle nose deformity and recurrent episodes of skin eruption associated with trauma over the knuckles, elbows and extensor surfaces of the arms, unresponsive to therapy and characterized by areas of well-demarcated erythema that evolved to bullae over a few days and healed with scarring and milia.

She needed admittance to the intensive care unit and endotracheal intubation. Flexible bronchoscopy obtained profuse haemorrhagic secretions from the tracheobronchial tree that led to visualization of an erythematous mucosa with extensive erosions, without clear active bleeding and lesions along the pharynx and larynx. Bronchoalveolar lavage ruled out alveolar bleeding as a specific acute inflammatory liquid was obtained. Its microbiological, immune and cellular study revealed normal or negative results. Extensive erosions, blistering, scarring and crusting of nasal mucosa, septal perforation and bilateral eustachian tube inflammation were demonstrated by flexible nasopharyngoscopy examination. Laboratory tests showed iron deficiency anaemia (haemoglobin 9.1 g/dl), elevated ESR (45 mm/h) and CRP (3.94 mg/dl). Immunology studies (extractable nuclear antigen, immunoglobulin count, antinuclear antibodies, antineutrophil cytoplasmic antibodies, anti-double-stranded DNA and anticardiolipin antibodies), porphyria, viruses and syphilis screen yielded normal or negative results. Imaging studies with chest X-ray and CT and echocardiography were normal. CT scan of the paranasal sinuses showed septal deviation, abundant secretions and mucosal thickening in maxillary and ethmoid sinuses, bilaterally.

Incisional biopsies were obtained from affected and healthy skin and serum was sent to an international reference centre. Biopsy and exudates of the conjunctiva and septal mucosa were performed. Histopathological examination revealed non-specific acute and chronic focal inflammatory infiltrates, with superficial ulcers at septal mucosa and affection of the adjacent epithelia at conjunctival mucosa. No evidence of eosinophils, signs and symptoms of systemic vasculitis or of a definite collagen vascular disease were identified.

of malignancy, granulomata, microorganisms nor any specific lesions were observed and cultures were negative.

The mucocutaneous lesions were considered suspicious of associated epidermolysis bullosa acquisita (EBA) [1], although this diagnosis could not be established at that moment, and just oculocutaneous, nasal, oral, pharyngeal, laryngeal and oesophageal mucosal involvement were previously reported [2]. While awaiting the skin results, a flare-up with nasal chondritis, eustachian tube affection and episcleritis led to the diagnosis of relapsing polychondritis (RP) [3, 4], and treatment was changed to pulse intravenous steroids (1 g/day for 3 days of 6-methylprednisolone) and cyclophosphamide (500 mg/week for 6 weeks and 1200 mg/month for 3 months) due to life-threatening involvement of Airways mucosa.

Histopathology of involved skin specimens revealed subepidermal bullae without eosinophils (Fig. 1), with linear staining of IgG at the basement membrane zone by direct immunofluorescence, supporting the diagnosis of EBA with severe tracheobronchial mucosal involvement. Treatment was changed to cyclosporin (5 mg/kg/day) due to the poor control of the haemoptysis and anaemia and the potential toxicity of cyclophosphamide, allowing progressive tapering of steroids and normalization of clinical and laboratory parameters, without significant side-effects. Indirect immunofluorescence of skin sections and of 1 M NaCl-split and immunoblotting of epidermal and dermal extracts (EBA antigen and anti-p200 pemphigoid antigen) were negative, but only 50–65% of these patients have circulating autoantibodies [5]. As cutaneous lesions cleared, immuno-electron microscopy analysis could not be performed.

After 1 yr, as steroids were discontinued, the patient developed a severe flare of CD with elevated ESR and CRP and recurrence of skin EBA, but refused further biopsies. As infliximab (4 mg/kg every 6 weeks) had proved effective for CD [6] and RP [7–9], it was added in combination with low-dose methotrexate, after latent TB was excluded. It led to remission and allowed cyclosporin dose reduction (3 mg/kg/day), methotrexate withdrawal to avoid toxicity derived from combined immuno-suppressive therapy and surgical reconstitution of entropium, nasal cartilage and septum, without complications. After 4 yrs of continuous treatment, clinical and laboratory parameters remain normal without new life-threatening flare-ups and significant side-effects.

We report an immune-mediated inflammatory syndrome that has not been described before, to our knowledge. We have not found mention of severe mucosal affection of middle and lower Airways due to EBA in literature. The cartilage matrix protein subdomain of the non-collagenous 1 domain is the first antigenic epitope on type VII collagen demonstrated to be a pathogenic target for EBA autoantibodies [10]. Although EBA is caused by autoantibodies against type VII collagen, present in skin and colon, it has been associated with other autoimmune diseases in which TNF-α plays a major role (supplementary data are available at Rheumatology Online). We may hypothesize that TNF-α could be involved in its pathogenesis and emphasize the successful use of infliximab for the management of a disease that remained particularly refractory to treatment.

Acknowledgement
We thank Dr Takashi Hashimoto, Department of Dermatology, Kurume University School of Medicine, Kurume, Japan, for the immunoblotting studies.

Disclosure statement: The authors have declared no conflicts of interest.

Supplementary data
Supplementary data are available at Rheumatology Online.

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Accepted 14 November 2007

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Advance Access publication 28 January 2008

Comment on: Atorvastatin therapy improves endothelial-dependent vasodilation in patients with systemic lupus erythematosus: an 8 week controlled trial

Sir, I have read with the interest the paper by Dr Ferreira and colleagues [1] regarding the therapeutic potential of atorvastatin in

Rheumatology key message

- This unreported immune-mediated inflammatory syndrome can be successfully managed with infliximab.