SIR, Pulmonary artery hypertension (PAH) is now the leading cause of mortality in patients with lcSSc. Here we report the case of a 29-yr-old woman with severe PAH as the presenting feature of ssSSc.

The patient first presented in March 2004 with a history of RP. There were no additional clinical features of CTD, in particular SSc. Digital thermography confirmed RP and nail-fold capillaroscopy demonstrated some elongated capillaries with punctate haemorrhages. Routine haematological and biochemical investigations were normal. She had a positive ANA, 1/2560 titre with nucleolar staining pattern on indirect IF. Further characterization of the ANA specificity was negative by routine methods, which at that stage did not include immunoprecipitation. A diagnosis of auto-immune RP was made and she was seen annually over the next 3 yrs with stable RP and no new symptoms or signs to suggest CTD.

In March 2007, she presented to hospital with worsening exertional dyspnoea, a dry cough and right-sided pleuritic chest pain. Her exercise tolerance subsequently declined significantly and she became breathless at rest (New York Heart Association, NYHA Grade 4). She had signs of right-sided heart failure with a raised jugular venous pressure and peripheral oedema. ECG revealed right bundle branch block and P-wave pulmonale suggestive of right atrial hypertrophy. A CT pulmonary angiogram (CTPA) was reported as showing some minor consolidation in the right middle lobe suggestive of right atrial hypertrophy. A CT pulmonary angiogram (CTPA) was reported as showing some minor consolidation in the right middle lobe and hypervascularity of right heart chambers but there was no evidence of pulmonary embolus or interstitial lung disease (ILD) (Fig. 1). Trans-thoracic echocardiography demonstrated high estimated right ventricular pressures with a tricuspid regurgitation gradient of 75 mmHg. On right heart catheterization, the patient had evidence of significant pulmonary hypertension, with a mean pulmonary artery pressure of 57 mmHg, cardiac index of 2.41/min/m² and a pulmonary vascular resistance of 11.9 WU.

On reassessment, there were no other clinical features of CTD, including no skin thickening. Her autoimmune serology was repeated, and a high-titre nucleolar ANA was demonstrated as previously. The specificity was further characterized using immunoprecipitation as being anti-U3RNP (fibrillarin) autoantibodies. Based on the finding of an SSc-specific autoantibody, and her clinical features, a diagnosis of ssSSc with isolated PAH was made. She was treated with diuretics, anticoagulants, and intravenous epoprostenol. She responded well to treatment and her

**Acknowledgements**

We would like to thank Dr Alex Keough for assistance with data collection at St George’s Healthcare NHS Trust and Dr Richard Morris, Department of Population Sciences, Royal Free and University College Hospitals Medical School for help with statistical analysis.

**Disclosure statement:** P.D.W.K. has received departmental support from Sanofi-Aventis. All other authors have declared no conflicts of interest.

A. KAUL1, D. T. O’REILLY2, R. K. SLACK2, D. COLLINS3, J. WALMSLEY4, O. DUKE4, P. D. W. KIELY1

1St George’s Healthcare NHS Trust, London, 2West Suffolk Hospital, Bury St Edmonds, 3Great Western Hospital, Swindon and 4Epsom and St Helier NHS Trust, Epsom, UK

Accepted 4 June 2008

Correspondence to: A. Kaul, Department of Rheumatology, Royal Free Hospital, Pond Street, London NW3 2QG, UK

E-mail: arvind.kaul@royalfree.nhs.uk

**Rheumatology key message**

- MTX and LEF combination therapy is well tolerated by a majority of patients.
exercise tolerance improved to Grade 2 NYHA symptoms (walking >200 yards at normal pace).

The first case report of clinical features of SSc in the absence of cutaneous manifestations described a patient with isolated gastrointestinal disease and subsequently the term ssSSc was defined [1,2]. In a study by Poormoghim et al. [3] of 48 patients with ssSSc, 98% had RP. Severe dyspnoea was more prevalent in the ssSSc cohort (P < 0.0004) compared with their lcSSc cohort, but it was not clear if this was due to ILD, ILD associated with PAH or primary PAH. However, primary PAH that they defined either clinically or by cardiac catheterization was more frequent in the ssSSc cohort (23%) compared with lcSSc cohort (13%) (P = NS). PAH was the leading cause of mortality in both groups but this was more frequent in the ssSSc patients (52% compared with 24%, P = 0.009). In addition, the most common autoantibody specificity in the ssSSc group was ACA and this is a recognized risk factor for PAH in SSc. Patients with ssSSc can present in other ways and there are reports of ssSSc renal crisis and ssSSc-associated ILD associated with other nuclear autoantibodies such as anti-RNAP-III and anti-Th/To [4–7].

To our knowledge this is the first published case of ssSSc presenting with isolated PAH associated with anti-U3 RNP autoantibodies. This autoantibody specificity has been previously reported in patients with dcSSc, lcSSc, overlap myositis, ILD, PAH and renal disease [8, 9]. Our case highlights the importance of close monitoring of patients who present with RP and a strongly positive nuclear autoantibody pattern, for further organ involvement such as PAH that can now be effectively treated if detected early enough. More widespread use of specialist techniques to further characterize such ANA patterns would help to identify those patients at particular risk. In addition, the presence of abnormal nail-fold capillaries at presentation can be helpful to identify those patients at particular risk. In addition, the most common autoantibody specificity in the ssSSc group was ACA and this is a recognized risk factor for PAH in SSc. Patients with ssSSc can present in other ways and there are reports of ssSSc renal crisis and ssSSc-associated ILD associated with other nuclear autoantibodies such as anti-RNAP-III and anti-Th/To [4–7].

Disclosure statement: H.G. is supported by the Arthritis Research Campaign. J.G.C. received research grants from GlaxoSmithKline. J.S. received travel grants from Enervys and Actelion. All other authors have declared no conflicts of interest.

J. D. Pauling1, H. Gunawardena1, J. G. Coghlan2, J. Easaw3, J. Suntharalingam4, N. J. McHugh4

1Royal National Hospital for Rheumatic Diseases, Bath, 2Department of Cardiology, Royal Free Hospital, London, 3Department of Cardiology and 4Department of Respiratory Medicine, Royal United Hospital, Bath, UK

Accepted 4 June 2008

Correspondence to: N. J. McHugh, Royal National Hospital for Rheumatic Diseases, Bath, BA1 1RL, UK. E-mail: neil.mcHugh@rnhd.nhs.uk

Rheumatology key message

- Screen patients with RP and scleroderma-specific auto-antibodies for pulmonary hypertension.

Rheumatology 2008;47:1432–1433
doi:10.1093/rheumatology/ken243
Advance Access publication 7 July 2008

Repairing erosions in rheumatoid arthritis. A realistic goal

Sir, Persistent inflammation in RA is an osteodestructive process, which leads to an accumulation of joint damage over time. Bone loss in RA occurs both in the joints and throughout the skeleton as a result of the multifactorial increase in bone resorption. Bone erosion starts early in disease and progresses most rapidly during the first year. The structural joint damage that is evident on conventional radiographs is strongly associated with poor functional outcome in patients with RA. These findings have fostered the concepts that retardation, arrest or even repair of structural damage should be considered central goals in the treatment of RA. We report a case of a woman with a long-RA patient, in whom erosion repair was achieved.

The patient was a 65-year-old woman who had been diagnosed with a positive RF RA in 1985. Since then, she had received multiple DMARDs (aurothiomalate, chloroquine, MTX, SSZ, HCQ, LEF and infliximab) without achieving a satisfactory clinical response. Radiographs taken during those years revealed disease progression, as illustrated by the serial images of her left first MCP joint (Fig. 1). In 2004, etanercept plus low-dose weekly MTX was initiated, obtaining an excellent clinical response, with the 28-joint disease activity score persistently under 3.2. Radiographs taken after 2 and 3 yrs with etanercept plus MTX showed repair of the erosions (Fig. 1).

RA results in extensive bone resorption as evidenced by focal bone erosions, juxta-articular osteopenia and systemic osteoporosis. Osteoclasts, specialized bone resorbing cells regulated by RANK ligand (RANKL) and monocyte colony-stimulating factor, are implicated in RA joint erosion [1]. Blockade of RANKL does not inhibit inflammation, but can prevent the structural damage progression that often occurs despite use of disease-modifying drugs, preserving joint architecture and function in RA patients. Anti-TNF agents (infliximab, etanercept and adalimumab) have shown the potential to arrest radiological damage in patients with active RA [2–4]. In this sense, the results from the Trial of Etanercept and Methotrexate with Radiographic Patient Outcomes (TEMPO) study suggest that etanercept in combination with MTX slows the radiographic progression in RA [5]. In this study, more patients receiving combination therapy had negative scores in the radiographic measurement after 1 and 2 yrs, compared with patients in either of the monotherapy groups, which could mean an effective repair of structural damage [5, 6]. The exact mechanism by which this protection occurs has not been fully determined, but it is thought that TNF-α antagonists modulate the osteoprotegerin (OPG)/RANKL system. In fact, increased expression of OPG, and subsequent decrease in the RANKL:OPG ratio, in synovial tissue have been seen in patients following therapy with an anti-TNF-α agent [7].

But the ambitious goal to repair bone erosions in RA could be achieved not only with the control of the inflammation, since new powerful agents are being developed to inhibit osteoclast