4. SACHS RN, TALVARD O, LANFRANCHI J: Myocarditis in adult Still's disease. *Int J Cardiol* 1990; 27: 377-80.
5. UEDA T, MIZUSHIGE K, SAKAMOTO S *et al.*: Adult Still's disease with myocardial dysfunction induced by microangiopathy. *Jpn Circ* 1997; 61: 74-7.
6. KAWASHIMA M, YAMAMURA M, TANIAI M *et al.*: Levels of interleukin-18 and its binding inhibitors in the blood circulation of patients with adult-onset Still's disease. *Arthritis Rheum* 2001; 44: 550-60.
7. KAWAGUCHI Y, TERAJIMA H, HARIGAI M, HARA M, KAMATANI N: Interleukin-18 as a novel diagnostic marker and indicator of severity in adult-onset Still's disease. *Arthritis Rheum* 2001; 44: 1716-8.
8. POMERANTZ BJ, REZIKOV LL, HARKEN AH, DINARELLO CA: Inhibition of caspase 1 reduces human myocardial ischemic dysfunction via inhibition of IL-18 and IL-1 β. *Proc Natl Acad Sci USA* 2001; 98: 2871-6.
9. MALLAT Z, HENRY P, FRESSONNET R *et al.*: Increased plasma concentrations of interleukin-18 in acute coronary syndromes. *Heart* 2002; 88: 467-9.
10. BLANKENBERG S, TIRET L, BICKEL C *et al.*: Interleukin-18 is a strong predictor of cardiovascular death in stable and unstable angina. *Circulation* 2002; 106: 24-30.

Fatal pulmonary hypertension in primary Sjøgren's syndrome

Sirs,

Pulmonary hypertension (PH) is a rare finding in Sjøgren's syndrome (SS) (1, 2). One year after primary SS was diagnosed in a 55-year-old woman (keratoconjuctivitis sicca, wrist/finger arthralgias, leukocytopenia (2.2 x 10^9/l), polyclonal increased immunoglobulins, high ESR (83 mm/hr), RF Latex-test 62 IU/ml and negative ANA), the patient developed intermittent, non-radiating chest pain unrelated to exercise with normal treadmill testing. Two years later (after an ineffective trial with hydroxychloroquin 400 mg daily), dyspnoea developed gradually with normal chest X-ray findings. Again 2 years later she was referred because of increasing dyspnoea. There was central cyanosis, increased jugular venous pressure, slight non-tender hepatomegaly without ascites or peripheral oedema, bilateral basal crackles, regular tachycardia (104/min) and fixed splitting of the second heart sound, drumstick fingers with normal skin and joint findings (no Raynaud's phenomenon).

Other findings: Schirmer < 10 mm, sialometry 0.1 ml (15 min), labial biopsy focus score 4.5 (27 infiltrates in a 24 mm^2 specimen), hypergammaglobulinemia (IgG 18.7 g/l IgM 2.75 g/l and IgA1.73 g/l), negative results for ANA (ELISA-screen), anti-SSA and -SSB (ELISA), RF and anticardiolipin antibodies and lupus anticoagulant and normal TSH/T4. Transthoracic ultrasound revealed a hypertrophied right ventricle, displacement of the interventricular septum into the left ventricle, normal pericardium and estimated systolic pulmonary artery pressure 90 mm Hg. Perfusion scan, spiral- and high resolution CT were normal, with FVC and FEV1 80% and 79% of predicted and DLCO 73%. At catheterisation, the right atrial pressure was 15 mm Hg, systolic pulmonary artery pressure 100 mm Hg (increasing to 115 mm Hg with static exertion) and capillary wedge pressure 11 mm Hg. The procedure had to be terminated prematurely due to hypotension. Despite treatment with oxygen (2–4 l/min), calcium antagonists, ACE-inhibitors, diuretics and low weight molecular weight heparin, periods of hypotensive syncope became increasingly frequent and within days led to refractory circulatory shock. Autopsy confirmed the right ventricular hypertrophy (right ventricle weight 117 gr, left ventricle 142 gr) but no other structural heart disease. There was no evidence of interstitial or thromboembolic lung disease and pulmonary parenchyma was normal, essentially with markedly thickened arterioles and severe proliferation of smooth muscle (Fig. 1), without evidence for vasculitis or IgG, IgM or C3 deposition.

Pulmonary hypertension (PH) is known to complicate a number of connective tissue diseases (2). As there was no evidence of scleroderma, SLE or mixed connective tissue disease and as she fulfilled European classification criteria (3), we consider pSS the most likely cause for the fatal PH in this patient.

The English literature contains nine cases of PH in SS patients. Thromboembolic pulmonary occlusion associated with antiphospholipid antibodies and a pulmonary form of Raynaud's phenomena were described as causes for PH, but both were absent in this patient (4). The main symptom of PH is dyspnoea, but this is a rather unspecific finding in pSS patients (5). Unresolved chest pains were also considered to be a non-specific pSS symptom in this case, but proved to be early signs of PH, where chest pain is the next most common symptom (6).

The prognosis for PH patients in general is dismal with a median survival of 2.8 years (2,7). While some beneficial effect of immunosuppressive treatment is described in the literature (8), PH treatment with vasoactive drugs (prostacyclin, endothelin-receptor antagonist) (9, 10) shows promising results. With more effective therapy available, the early detection of PH with screening by non-invasive cardiac ultrasound becomes crucial, as illustrated also in pSS patients with unexplained dyspnoea and/or chest pains.

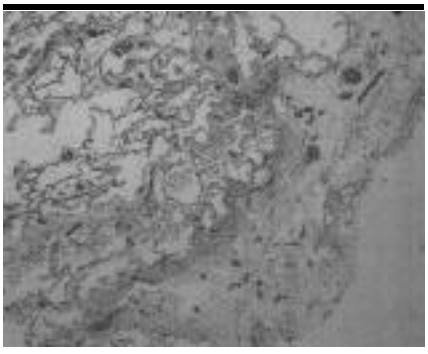

Fig. 1. Haematoxylin-eosin staining of pulmonary tissue showing largely intact lung parenchyma with spreaded lymphocyte infiltration, severe smooth muscle hypertrophy obliterating the vasculature without signs of vasculitis or trombi (**arrow**). (Courtesy of Dr. Tor Arne Hanssen, Dept. of Pathology, University Hospital North Norway).

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References

1. CAIN HC, NOBLE PW, MATTHAY RA: Pulmonary manifestations of Sjogren's syndrome. *Clin Chest Med* 1998; 19: 687-99.
2. MAGLIANO M, ISENBERG DA, HILLSON J: Pulmonary hypertension in autoimmune rheumatic diseases: where are we now ? *Arthritis Rheum* 2002; 46: 1997-2009.
3. VITALI C, MOUTSOPOULOS HM, BOMBARDIERI S: The European Community Study Group on diagnostic criteria for Sjögren's syndrome. Sensitivity and specificity of tests for ocular and oral involvement in Sjogren's syndrome. *Ann Rheum Dis* 1994; 53: 637-47.
4. GALLERANI M, GOVONI M, RICCI L *et al.*: a 49-year-old woman with dyspnoea, palpitations and syncope. *Int J Cardiol* 1996; 55: 67-78.
5. DAVIDSON BK, KELLY CA, GRIFFITHS ID: Ten year follow up of pulmonary function in patients with primary Sjogren's syndrome. *Ann Rheum Dis* 2000; 59: 709-12.
6. RASHID A, LEHRMAN S, ROMANO P, FRISHMAN W, DOBKIN J, REICHEL J: Primary pulmonary hypertension. *Heart Dis* 2000; 2: 422-30.
7. McLAUGHLIN VV: Medical management of primary pulmonary hypertension. *Expert Opin Pharmacother* 2002; 3: 159-65.
8. MARIETTE X, BRENOT F, BROUETJC: Recovery from pulmonary hypertension with steroid therapy in a patient with Sjögren's syndrome and polymyositis. *J Rheumatol* 1994; 21: 772-3.
9. CHANNICK RN, RUBINLJ: New and experimental therapies for pulmonary hypertension. *Clin Chest Med* 2001; 22: 539-45.
10. CHANNICK RN, SIMONNEAU G, SITBON O *et al.*: Effects of the dual endothelin-receptor antagonist bosentan in patients with pulmonary hypertension: a randomised placebo-controlled study. *Lancet* 2001; 358: 119-23.

Myositis as a presenting feature of polyarteritis nodosa

Sirs,

Polyarteritis (PAN) is a vasculitis affecting predominantly the small and medium-size arteries (1); less commonly it may affect the muscles but this is not a frequent nor a main feature of the disease. We report a patient admitted to our hospital with a clinical pic-