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.

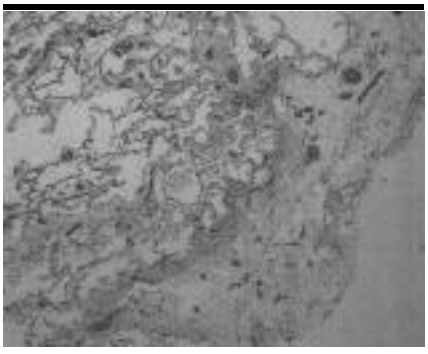

Fig. 1. Haematoxylin-eosin staining of pulmonary tissue showing largely intact lung parenchyma with spreaded lymphocyte infiltration, severe smooth muscle hypertrophy obliterating the vasculature without signs of vasculitis or trombi (**arrow**). (Courtesy of Dr. Tor Arne Hanssen, Dept. of Pathology,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

H.K. Aslaksen[1] H.C. Nossent[1,2]

[1]University Hospital Northern Norway, Department of Rheumatology, Tromsø; [2]University of Tromsø Medical School, Institute of Clinical Medicine, Department of Rheumatology, Tromsø, Norway.

Address for correspondence: Hans C. Nossent, MD, Department of Rheumatology, University Hospital Northern Norway, N-9038 Tromsø, Norway. E-mail: hans.nossent@unn.no

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-

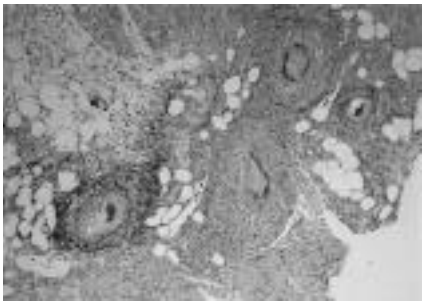

Fig. 1. Weigert e.f., 10X: quadriceps muscle biopsy showing transmural infiltration of lymphocytes and neutrophils in a small-sized artery with fibrinoid necrosis, partial destruction of the internal lamina elastica and lumen occlusion.

ture of paraplegia secondary to severe myopathy as the presenting and only manifestation of polyarteritis.

A 68-year-old man was referred to our department with a 2-month history of fever and progressive weakness mainly in the lower limbs, to the extent that he was unable to rise from his bed or to walk. Apart from chronic hepatitis due to HBV he had no other illnesses and his past medical history was unrevealing. On examination he was pyrexial (39.6°C), with no rashes or palpable lymph nodes. His blood pressure was 110/80 mmHg, he had normal heart sounds and all peripheral pulses were present. He presented generalised weakness that was quite severe in the proximal muscles of the lower limbs, with marked wasting and tenderness of the thigh muscles. His deep tendon reflexes were absent but sensibility was preserved.

Tests revealed a erythrocyte sedimentation rate (ESR) of >100 mm/hour, ferritin > 1000 μgr/L, creatin kinase (CK) at 890 IU (n.v.< 200), and leucocytosis (22,970 cell/mm^3) with absolute neutrophilia (19,500 cells/mm^3). Antinuclear antibodies (ANA), anti-neutrophil cytoplasmic antibodies (ANCA), rheumatoid factor (RF), cryoglobulin, HCV Ab were all negative, but HbSAg was positive. Renal function was normal.

During his admission the patient underwent an electromyography that showed a myopathic pattern with polyphasic potentials of short duration and low amplitude. A biopsy of his quadriceps muscle biopsy revealed a transmural infiltration of lymphocytes and neutrophils in small size arteries with fibrinoid necrosis, disruption of the lamina elastica and lumen occlusion (Fig 1).

A diagnosis of PAN was made and treatment was started with prednisone 1 mg/kg/die in divided doses. No other immunosuppressive drug was added because proteinuria >1 gr/die, cardiomyopathy, gastrointestinal and central nervous system involvement were absent. Agradual recovery was observed, after 2 weeks the fever had subsided, ESR was 40/hour and he was able to move fully against gravity but not resistence and he was discharged on the same therapy.

Our patient presented with severe muscle atrophy, tenderness and weakness mimicking a polymyositis (2). This presentation, together with elevated levels of CK highly suggested a myositis, but this diagnosis was ruled out by the histopathology of the muscle biopsy which were consistent with PAN. This represents a rare case of PAN presenting as myositis. Forms of PAN limited to muscle involvement without generalized weakness and systemic features have been reported, but were considered by some to be a distinct variant of PAN (3). Cases of systemic disease (fever, weight loss), and generalized myopathy are less frequently reported and to best of our knowledge this is only the fourth case reported.

Fort *et al*. (4) reviewed 7 reported patient series of PAN and found that muscular symptoms were present in 51% of cases but clinical myopathy as a presenting feature was uncommon. In the same paper the authors reported a patient with a clinical picture of polymyositis and elevated CK but a normal muscle biopsy. The diagnosis of PAN was made by histopathology of the colonic mucosa. Plumley *et al*. (5) and Sumer (6) have reported one case each; in both patients the diagnosis was made by muscle biopsy and in the latter case with a normal EMG.

In conclusion our case offers two interesting observations: first, PAN can present as a myopathy; second, in a patient with clinical myopathy the diagnosis of PM requires a muscle biopsy even in cases such as the one described here, where clinical, laboratory and EMG findings suggest the diagnosis of PM.

S. LUPOLI[1], *MD* R. ROSSI[2], *MD*
A. GARGIULO[1], *MD* P.R.J. AMES[3], *MD*

[1]*Division of Internal Medicine, Unit of Rheumatology and* [2]*Division of Pathology, Ospedale S. Sebastiano, Caserta, Italy;*
[3]*Academic Department of Rheumatology , Leeds General Infirmary Leeds, UK.*

Correspondence to: Dr Salvatore Lupoli, Unit of Rheumatology, Division of Internal Medicine, Ospedale S. Sebastiano, via Tescione, 81100 Caserta, Italy.
E-mail: Salvatore_lupoli @ tin.it

References

1. LIGHTFOOT RWJR: Overview of the inflammatory vascular diseases. *In* KLIPPEL JH and DIEPPE PA (Eds): *Rheumatology*, 2nd edition, Mosby Ltd, 1998, 7,17:1-6.
2. MASTAGLIA FL, PHILLIPS BA: Idiopathic inflammatory myopathies: epidemiology, classification, and diagnostic criteria. *Rheum Dis Clin North Am* 2002; 28: 723-41.
3. NAKAUCHI Y, SUEHIRO T, KUMON Y *et al*.: Localized polyarteritis of the forearm and epididymis. *Intern Med* 1994; 33: 48-52.
4. FORT JG, GRIFFIN R, TAHMOUSH A, ABRUZZO JL: Muscle involvement in polyarteritis nodosa: Report of a patient presenting clinically as polymyositis and review of the literature. *J Rheumatol* 1994; 21: 945-8.
5. SUMER M, OKSUZOGLU B, SUNGUR C, GUNGEN Y: Muscle involvement in polyarteritis nodosa: Report of a case presenting clinically as myopathy. *Clin Exp Rheumatol* 2002; 20: 434.
6. PLUMLEYSG, RUBIO R, ALASFAR S, JASIN HE: Polyarteritis nodosa presenting as polymyositis. *Semin Arthritis Rheum* 2002; 31: 377-83.