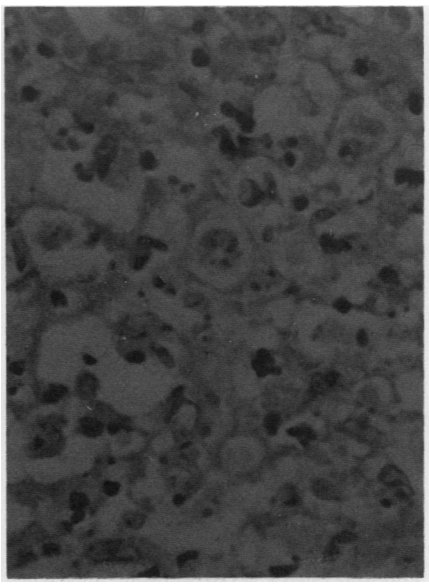
LETTERS TO THE EDITOR

'Lupus lymphadenitis' simulating a strangulated femoral hernia in a patient with mixed connective tissue disease

Sir: Lymphadenopathy is a common finding in both systemic lupus erythematosus (SLE) and mixed connective tissue disease (MCTD)[1] and is often associated with disease activity. Although lymph nodes may be tender and the degree of lymphadenopathy sufficient to cause concern over the possibility of primary lymphatic disease,[2] an acute presentation with localised lymphadenopathy in the absence of other systemic features is exceptional. We report a patient in whom the clinical presentation suggested the diagnosis of a strangulated femoral hernia, but in whom the true diagnosis, proved histologically, was lupus lymphadenitis.

A 25 year old woman presented with a five day history of a painful lump in her right groin associated with a raised temperature and general malaise. She had had some transient swelling in her right groin five months earlier. In 1983 she had had a partial thyroidectomy for thyrotoxicosis and in 1986 she was diagnosed as having SLE when she presented with general malaise, weight loss, Raynaud's phenomenon, and a cough. She was found to have diffuse hypergammaglobulinaemia, a negative rheumatoid factor, a positive antinuclear factor (titre 1/80, speckled pattern), positive anti-DNA binding (titre 1/2560), and a positive extractable nuclear antigen (titre 1/732). She then had two episodes of myopericarditis, both of which responded well to intravenous methylprednisolone. She has now developed sclerodermatous changes affecting her hands and face and oesophageal dysmotility. She has never had any clinical features to suggest Sjögren's syndrome. Her autoantibody pattern has changed with the development of antiribonucleoprotein antibodies and the loss of DNA binding, suggesting progression to MCTD with features of scleroderma. Throughout this time her maintenance dose of oral prednisolone and azathioprine has been continued. On examination she was cushingoid with facial telangiectasia, microstomia, and sclerodermatous changes in her hands. She was afebrile, but had a tender mass in her right groin measuring 3×3 cm, which was thought to be a strangulated femoral hernia. At operation she was found to have several enlarged lymph nodes below the right inguinal ligament. She made a good postoperative recovery and has since remained well with her usual maintenance dose of prednisolone 7 mg/day and azathioprine 50 mg/day. Histology of the lymph nodes was characteristic of lupus lymphadenitis showing necrosis (figure) with nuclear debris undergoing phagocytosis in one lymph node and both centroblastic and immunoblastic responses in adjacent nodes.

Localised or generalised lymphadenopathy occurs in about half of patients with SLE at some stage during their disease[1] and in about a third of patients with MCTD.[3] In SLE superficial nodes, particularly those in the neck, are commonly affected.[1,4] Necropsy studies, however, commonly show enlargement of mesenteric, tracheobronchial, and retroperitoneal



Section of lymph node showing marked necrosis. (Haematoxylin and eosin.)

nodes, suggesting more generalised involvement. The pathology of lymph nodes in SLE is well reported and characteristic,[1,4–6] though the pathogenesis remains unknown.[1] Affected lymph nodes show a necrotising lymphadenitis with lymphoblastic and immunoblastic responses. Advanced lesions may also show haematoxylin bodies and deposits of haematoxyphilic material in the walls of blood vessels (the Azzopardi phenomenon) with perivascular inflammation.[7]

There are few reports of the pathology of lymph nodes in MCTD. Most are said to show non-specific changes.[1] Two cases have been described, however, in which necrosis was associated with lymphoid hyperplasia.[8] In one of these the necrosis was associated with thrombosis, resulting in hilar infarction.

Other types of lymphadenopathy may also be found in patients with connective tissue disease. Malignancy may occur more commonly in patients with SLE, particularly those receiving immunosuppressive drugs.[9] Lymphoproliferative disorders have been reported to be associated with an increased incidence of rheumatoid arthritis compared with a control population[10] and to occur 44 times more commonly in patients with Sjögren's syndrome.[11] Furthermore, angioimmunoblastic lymphadenopathy, a non-neoplastic proliferative disorder of lymphocytes, shows many clinical and laboratory similarities to SLE and may be associated with it.[12]

In conclusion, we know of no previous report of lupus lymphadenitis simulating a surgical emergency. This case also underlines the fact that no clear pathological distinction can be drawn between SLE and MCTD.

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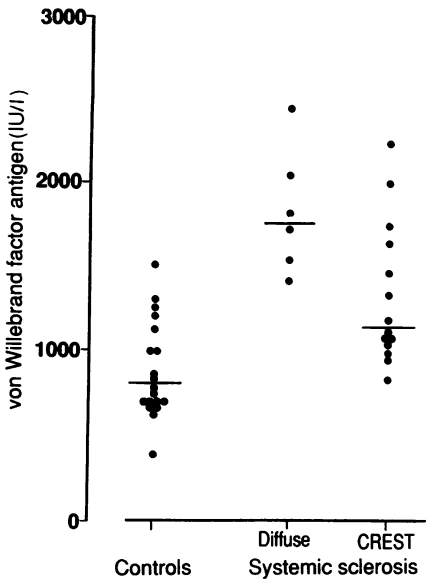
Raised concentrations of von Willebrand factor antigen in systemic sclerosis

Sir: von Willebrand factor antigen is the antigenic component of von Willebrand factor, and circulating concentrations are raised in vasculitis and systemic sclerosis, reflecting damage to endothelial cells.[1,2] In systemic sclerosis concentrations of von Willebrand factor antigen correlate with a risk of mortality[3] and with the extent of visceral disease—that is, involvement of heart, lung, muscle, etc.[4] Neither of these reports directly considered the question of concentrations of von Willebrand factor antigen in the two major subgroups of systemic sclerosis—the limited cutaneous variant (CREST) and the diffuse variant.

We therefore studied 14 patients with clearly defined CREST but no proximal skin disease and six with the clearly defined diffuse variant with proximal scleroderma,[5] and compared them with 20 apparently healthy laboratory and hospital staff as controls matched for age and sex. von Willebrand factor antigen in plasma was measured by a standard enzyme linked immunosorbent assay (ELISA).[6] Results are shown in the figure. von Willebrand factor antigen was raised in patients with systemic sclerosis relative to normal controls (median 1500 IU/l, controls 800 IU/l, $p<0{\cdot}01$, Mann-Whitney U test used throughout). Analysis by subgroup showed that both the diffuse variant (1770 IU/l, $p<0{\cdot}01$) and the CREST variant (1150 IU/l, $p<0{\cdot}05$) had raised concentrations of von Willebrand factor antigen, which were higher in the patients with the diffuse variant than in those with the CREST variant ($p<0{\cdot}01$).

These data confirm and extend previous reports about von Willebrand factor antigen in systemic sclerosis. Sheeran *et al* studied 34 patients with systemic sclerosis, of whom seven died (mean von Willebrand factor antigen 2980 IU/l, in the surviving group 1450 IU/l).[3] Greaves *et al* found a mean von Willebrand factor antigen of 1730 U/l in 16 patients with severe disease and 1030 U/l in 12 patients with mild disease.[4] Our finding



Plasma von Willebrand factor antigen concentrations. Each point represents one patient or control. The bar is the median value.

of a higher von Willebrand factor antigen in patients with diffuse disease suggests endothelial cell damage may be more extensive and active in this group. The extent of vascular injury as shown by von Willebrand factor antigen concentrations may be regarded as a prognostic marker in these disorders.

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Anticardiolipin antibodies in drug addicted patients with AIDS

Sir: The presence of anticardiolipin antibodies and lupus anticoagulant has been described in up to 90% of homosexual patients with AIDS.[1 2] There have been some attempts to relate anticardiolipin antibodies with *Pneumocystis carinii* infection,[3] other opportunistic infections, and with neoplasm,[4] but up to the present, anticardiolipin antibodies in patients with AIDS have only been associated with a poor outcome of the illness.[5 6] In these patients anticardiolipin antibodies do not have the same clinical significance as they have in systemic lupus erythematosus.[1]

As drug addicted patients with AIDS have a different spectrum of rheumatic manifestations, with less reactive arthritis, Reiter's syndrome, and HIV related arthritis, and more septic arthritis than homosexual patients with AIDS (Monteagudo I *et al*, unpublished data), we studied the presence of anticardiolipin antibody in drug addicted patients with AIDS and its relation with clinical manifestations.

Anticardiolipin antibody was determined (Cheshire Diagnostic QACA enzyme linked immunosorbent assay (ELISA) kit) in 55 drug addicted patients. Forty three of these had been admitted to hospital because of acute problems related to AIDS (mainly infections) and the remaining 12, without acute problems, were seen at the outpatient AIDS clinic in the hospital. Anticardiolipin antibodies were positive in 42 (76%)—31/43 (72%) of the hospital patients and 11/12 (92%) of the outpatients. Clinical problems in the hospital patients were diverse, and we found no significant relation between infection and the presence of anticardiolipin antibodies. Additionally, there was no relation between anticardiolipin antibodies and the antecedent and kind of infection in these patients. Of the 55 patients, 43 were in stage IV AIDS, with 34 (79%) being positive for anticardiolipin antibodies. The remainder were in other stages and anticardiolipin antibodies were found in 10/12 (83%).

Our results show that the presence of anticardiolipin antibodies in drug addicted patients with AIDS is similar to that found in homosexual patients with AIDS. We found no relation between anticardiolipin antibodies and clinical manifestations (mainly infections) nor pathological antecedents in these patients. Similarly, there were no differences between outpatients and hospital patients. As with homosexual patients with AIDS, we found no correlation between anticardiolipin antibodies and recurrent thrombosis and thrombopenia. Probably, anticardiolipin antibodies are associated with HIV infection itself or with an abnormal immune response which is not yet well defined.

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Disabling ossification of the patellar tendon

Sir: A 42 year old lorry driver without any medical or traumatic history had had for two years pains in the right knee when walking. They had appeared simultaneously with the use of a new truck, in which the accelerator pedal was particularly stiff. After some months of limping clinical examination showed that the right patellar tendon was diffusely thickened and tender; a bulge sign was found, as well as atrophy of the quadriceps muscle and a marked reduction of the knee flexion. Erythrocyte sedimentation rate was 1 mm/1 st h; fasting blood sugar, calcium, phosphorus, and alkaline phosphatase were normal. Synovial fluid contained $0{\cdot}1\times10^9$ leucocytes/l without crystals.

Lateral plain radiographs and tomograms (without signs of patella alta: length of the patellar tendon equal to the diagonal length of the patella[1]) showed ossification of the tendon, which did not affect its distal third (fig 1). Sonography showed that ossification was mostly in the lateral part and that the non-ossified distal third was thickened in comparison with the tendon on the left side (anteroposterior thickness 9 mm *v* 6 mm). Radiographs and computed tomography also showed irregular enthesitic osteophytes of the upper part of patellae on both knees and, in addition, a femoral and tibial osteopoikilosis. Radiographs of cervical, thoracic, and lumbar spine showed no abnormalities.

Arthroscopic examination showed the joint cavity was normal. During operation the orthopaedic surgeon found that the right patellar tendon was wider than normal. The ossified mass was not adherent to the adjacent patella or tibia and could be easily dissected and removed. The patient recovered slowly and was able to resume his work three months later.

Pathological examination showed compact remodelled lamellar bone. In the mass of the bone, and on the proximal end, strips of fibrous cartilage were seen, which might be considered as an abnormal metaplasia of the tendon (figs 2A and B). Despite its distance from any insertion site this intermingling of bone and fibrous cartilage resembles that seen in enthesopathic hyperostosis—that is, in a location in which fibrocartilaginous bundles are normally found.[2] On the distal end tendinous bundles intermingled with scar tissue associated with some degree of scar

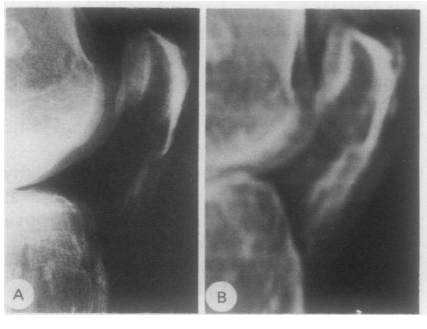

Figure 1 (A) Plain radiograph; (B) tomogram.