A 51-year-old woman complained of persisting fever, malaise, and myalgia in October, 2000. She was admitted to hospital in December, 2000, and had tests for fever of unknown origin: a full blood count, serum urea and electrolytes, blood and urine cultures, serological tests for antibodies to Brucella spp, syphilis, Coxiella burnetti, toxoplasmosis, HIV, hepatitis C and B, and cytomegalovirus. A Mantoux test, chest radiography abdominal ultrasound, echocardiogram, thoracic and abdominal computed tomography showed no abnormalities. A 67 Gallium full-body scan showed increased pulmonary uptake. She was given a presumptive diagnosis of sarcoidosis, but was not treated, as her fever resolved spontaneously. In January, 2001, the fever returned and she developed leg pain and weakness, and was admitted to our hospital.

Light microscopy of muscles biopsy specimen
(A) Neoplastic lymphoid cells in a small artery of skeletal muscle
(B) CD20 positive surface of intravascular cells

She was febrile and had generalized oedema. On neurological examination we found weakness and areflexia of both legs with bilaterally diminished pin-prick sensation. She had an erythrocyte sedimentation rate of 91mm, lactate dehydrogenase 1655 U/L, albumin 271 g/L, and C reactive protein 114 mg/L. We repeated the previous set of tests and did spinal radiographs, computed tomography, and magnetic resonance imaging. We did a lumbar puncture and analysed the cerebrospinal fluid, but could find no significant abnormalities. Electromyography showed lumbosacral polyradiculopathy, most prominent at the right S1 segment. The patient developed anasarca and ascending paraparesis with sacral and autonomic involvement. She became comatose and was transferred to the intensive care unit. A muscle biopsy showed intravascular occlusion by neoplastic lymphoid cells with large and hyperchromatic nuclei. These cells where strongly positive for CD20 antibody indication a B-cell lineage. We started chemotherapy with cyclophosphamide, vincristine, adriamycin, and prednisolone. The coma and anasarca resolved. Complete remission was achieved after six courses of biweek therapy. We did an autologous bonemarrow transplant in July, 2001, and when last seen in January, 2002, she could walk alone with a stick. Although she still had radicular right leg pain.

Neoplasia accounts for 20% of fevers of unknown origin, and a half of these are to lymphoma.1

Intravascular lymphoma is a rare variant of non-Hodgkin lymphoma, in which large neoplastic lymphocytes occlude the small vasculature. Selective intravascular growth may be due to lack of expression of CD29 and CD54 molecules in neoplastic lymphocytes.2 Neurological signs may cause multifocal cerebrovascular events, subacute encephalopathy, spinal cord and root symptoms, and peripheral or cranial neuropathies.3 Our patients polyradiculopathy developed into a subacute conus medullaris and cauda equina syndrome. Anasarca reported in this case is an unusual symptoms of non-Hodgkin lymphoma 5 and intravascular lymphoma, caused by vascular or lymphatic occlusion.
is confirmed by biopsy of symptomatic or swollen tissues. Muscle biopsy seems a safe and useful alternative in intravascular lymphoma with neurological signs.

References