

A young woman with generalised lymphadenopathy

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CASE REPORT

A 25-year-old woman of Dutch origin was referred to the outpatient clinic of a university medical centre because of generalised lymphadenopathy, anaemia, and thrombocytopenia. The patient had been healthy until three months ago. She had visited her general practitioner because of a red and painful eye. She insisted on a blood analysis, which revealed an abnormal blood count. There were no complaints of fever, weight loss, bleeding diathesis, arthralgias, exanthema or night sweats. She had no relevant medical history. Her eye complaints disappeared spontaneously after a few days.

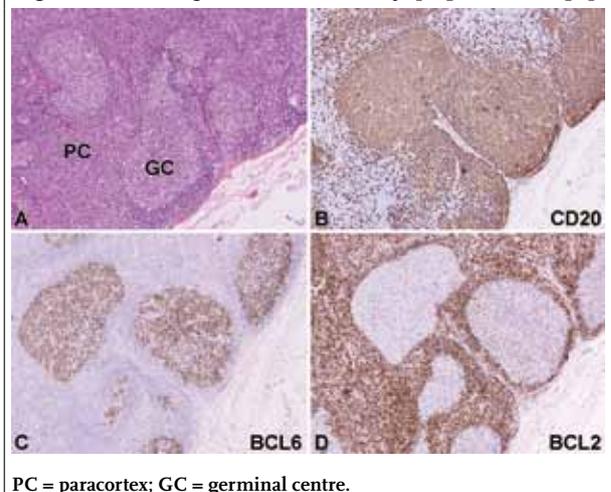
Physical examination showed submandibular, axillary and inguinal bilateral lymphadenopathy, with elastic, small (<1 cm) and non-tender nodes. There was enlargement of the spleen. The body temperature was 37.0 °C. The remainder of the examination was unremarkable.

Laboratory investigations revealed: erythrocyte sedimentation rate 83 mm/U (0 to 20 mm/U), haemoglobin 6.3 mmol/l (7.5 to 10 mmol/l), mean corpuscular volume 86 fl (80 to 100 fl), white blood cells normal, platelets $36 \times 10^9/l$

Figure 1. PET scan showing pathological FDG accumulation in multiple lymph nodes both above and below the diaphragm, and splenomegaly with diffuse increase of FDG accumulation



Figure 2. Histological examination of lymph node biopsy.



PC = paracortex; GC = germinal centre.

(A) Detail of a lymph node showing follicular hyperplasia and expansion of the paracortical zone. Haematoxylin and eosin stain. (B)-(D) Immunohistochemistry demonstrating the non-neoplastic nature of the lymph node. (B) B-lymphocytes showing membrane expression of CD20. (C) Germinal centre B-lymphocytes showing nuclear expression of the germinal-centre specific transcription factor protein BCL-6. (D) T- and B-lymphocytes showing cytoplasmic expression of the antiapoptotic protein BCL-2. Germinal centre B cells typically lack BCL-2 expression. Magnification x 40.

(150 to $400 \times 10^9/l$), creatinine 75 $\mu\text{mol/l}$ (65 to 95 $\mu\text{mol/l}$), and lactate dehydrogenase 216 U/l (0 to 247 U/l). Virus serology including Epstein-Barr virus, cytomegalovirus and human immunodeficiency virus was negative.

An FDG-PET scan showed pathological FDG accumulation in multiple lymph nodes both above and below the diaphragm, and splenomegaly with diffusely increased FDG accumulation (figure 1). In addition, a lymph node was excised (figure 2).

WHAT IS YOUR DIAGNOSIS?

See page 288 for the answer to this photo quiz.

When the history taking was repeated the patient stated she had photosensitivity. Additional laboratory findings revealed: direct Coombs reaction 3+, positive antinuclear antibody, positive antidouble stranded DNA and active urinary sediment (microalbumin/creatinine ratio 96.55 mg/mmol, erythrocytes 92 /ul, leucocytes 14 /ul). The lymph node revealed a *reactive lymphadenopathy*, without signs of malignancy. In the lymph node, neither immunophenotyping nor B cell receptor gene rearrangement analysis could demonstrate a clonal B cell proliferation.

Altogether, SLE was diagnosed based on the following criteria: photosensitivity, haematological and renal disorders, and positive serological testing. Extensive lymphadenopathy, as seen in this case, is a rare first presentation of SLE.¹

In the evaluation of Hodgkin's and non-Hodgkin's lymphoma (NHL) a PET scan with 2-deoxy-2-[¹⁸F]fluoro-D-glucose (FDG) has been used as a valuable diagnostic tool. In contrast, in SLE, FDG-PET has so far solely been used in the evaluation of central nervous system involvement.² FDG uptake is not tumour specific for malignancies because inflammatory lesions can also concentrate the tracer, including infectious diseases, inflammatory conditions such as vasculitis and arthritis, and granulomatous diseases such as tuberculosis and sarcoidosis.³ Our case shows that it is important for a physician to always keep an open eye for an alternative diagnosis. The more widespread use of FDG-PET scanning may more often lead to the suspicion of lymphoma in benign conditions such as autoimmune diseases.

Histopathology remains the golden standard method for the diagnosis and classification of persistently enlarged

lymph nodes. In SLE, the lesion is characterised by varying degrees of coagulative necrosis with haematoxylin bodies or reactive hyperplasia.⁴

In summary, SLE can present with extensive lymphadenopathy. Furthermore, the pattern of FDG uptake on a PET scan in SLE can mimic lymphoma. Being aware of the similarities between SLE and lymphoma is of the utmost importance for making the correct diagnosis.

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