

key points

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Systemic lupus erythematosus (SLE or lupus) is a multisystem, autoimmune disease. It can present in women or men at any age but is most common in women of reproductive age, especially those of African-Caribbean or South Asian descent. Lupus affected nearly 1 in 1,000 of the UK population in 2012 with a mean age at diagnosis of 49 years. Lupus nephritis occurs in a third of lupus patients in the UK and is associated with significant mortality and morbidity.

Lupus mainly affects women and causes fatigue, rashes (often associated with photosensitivity), hair loss and inflammatory arthritis. However, lupus can also affect men in whom it is often more severe with a greater likelihood of renal involvement and a greater degree of organ damage. Constitutional (e.g. fever and lymphadenopathy), cardio-respiratory (e.g. pleurisy and pericarditis), gastrointestinal (e.g. gall bladder vasculitis) and hepatic (e.g. autoimmune hepatitis) features may occur in up to 60% of patients. Eye involvement, other than sicca symptoms, is rare but potentially sight threatening and requires prompt referral to an experienced ophthalmologist.

An autoantibody screen should be performed when there is a clinical suspicion of SLE. Around 95% of lupus patients are ANA positive. ANA is not specific for lupus and may occur in other conditions including Sjögren's syndrome, systemic sclerosis, dermatomyositis, viral infections such as infectious mononucleosis and cancer. Lupus patients should be tested for antiphospholipid antibodies as these confer an increased risk of arterial/venous thrombosis and adverse pregnancy outcomes.

A low CRP with a raised ESR is typical of active lupus unless infection or effusions are present. According to the BSR guideline, the diagnosis of SLE requires a combination of clinical features and the presence of at least one relevant immunological abnormality. Biopsy proven lupus nephritis in the presence of ANA or anti-dsDNA antibodies may also be classified as lupus without the need for other criteria. Disease activity is categorised as mild, moderate or severe, and worsening disease activity is termed a flare.

Patients with lupus should be monitored regularly for disease features, drug toxicity and comorbidities. Those with active disease should be reviewed at least every 1-3 months with blood pressure, urinalysis, renal function, liver function tests, anti-dsDNA antibodies, complement levels and full blood count, with further tests as necessary. In active lupus, the ESR is usually high and CRP low, so an increase in CRP should prompt an infection screen. Those with stable low disease activity or in remission may be reviewed less frequently, such as 6 to 12 monthly. Patients with lupus are at increased risk of infection, atherosclerotic disease, malignancy, osteoporosis and avascular necrosis. They should be screened at least annually for risk factors including hypertension, dyslipidaemia, diabetes, high BMI and smoking.