Systemic Lupus Erythematosus

Heterogeneous, inflammatory, multi-system, idiopathic autoimmune dz with antinuclear Abs

Epidemiology

- Prevalence is 50-100 cases per 100,000.
- Peak onset age is 20-35. 9F:1M
- More common in Asian, Aborigine and Afro-Caribbean origin.

Risk Factors

- Genetics: HLA types DR3 and DR2, those with defective complement genes.
- Environmental: UV light, viruses, e.g. EBV
- Drugs: chlorpromazine, methyldopa, hydralazine, isoniazid, penicillamine & minocycline.

Presentation

Remitting and relapsing illness. Any major organ involvement usually<5yr of onset.
Non-specific: fatigue, malaise, fever, weight loss
Arthralgia: Peripheral, symmetrical, flitting Polyarthritis. Early morning stiffness but rarely swelling. Occ joint deformity and subluxation (Jaccoud's arthropathy). Fibromyalgia common.
Mucocutaneous: Photosensitivity rash – classically malar (butterfly) or discoid. Also livedo reticularis, patchy non-scarring alopecia, vasculitic rashes & mouth ulcers. Raynaud's in 20%
Renal: Glomerulonephritis common.
Cardiovascular: pericarditis, HT, Libman-Sacks endocarditis, ↑IHD
Pulmonary: pleurisy, fibrosing alveolitis, PE risk (if have antiphospholipid syndrome)
Neuropsychiatric: anxiety, depression, psychosis, seizures, neuropathy, meningitis.
Neurological: varied incl CVA due to vasculitis or thrombosis (antiphospholipid syndrome).
RES: Splenomegaly & lymphadenopathy

Investigations

Urine: UA (blood, protein - nephritis)
Blood: FBC (mild normochromic anaemia), ESR↑ (CRP may be normal), UEC (renal fn)
Serology: ANA (sens not spec), anti-dsDNA (spec not sens), other nuclear Abs (Ro, La, RNP & Sm often), antiphospholipid antibodies (anti-cardiolipin & lupus anticoagulant assoc with antiphospholipid syndrome), C3↑, C4↑
Other (organ involved): ECG/Echo (heart), CXR (lung), CT/MRI (brain), renal biopsy (kidney)

Management

Supportive: Counselling, ↓sun exposure, flu vaccination, stop precipitating drug. Pregnancy Cx (recurrent miscarriage, PET, IUGR, prematurity, thrombosis) may need LMW heparin, aspirin
Simple analgesics: NSAIDs used with caution because of GIT, renal, and CVS risks.
Treat any organ complications.
Hydroxychloroquine: 1st line in mild SLE (mucocutaneous lesions, arthralgia) **SE:** occulotoxic.
Plasma exchange: holding measure until the immunosuppressive therapy takes effect.
Immunosuppression (moderate-severe disease): **corticosteroids**, **cyclophosphamide** **azathioprine**, **methotrexate** and **cyclosporin**. IV **gammaglobulin** and **GCSF** if plts↓ and WCC↓.

Antiphospholipid (Hughes') syndrome

20-35% of SLE patients. Characterised by lupus anticoagulant ± cardiolipin antibodies, arterial/venous thrombosis, recurrent fetal loss & thrombocytopenia. Rx: aspirin or warfarin.

Prognosis

5yr survival rate >90%. ↑Morbidity & mortality with renal or multisystem involvement.