TIPS & PEARLS

SLE

SYSTEMIC LUPUS ERYTHEMATOSUS

- Multisystem
- Autoimmune
- Autoantibodies are made against a variety of autoantigens which form immune complexes
- Inadequate clearance results in a host of immune responses which cause tissue inflammation and damage

- Butterfly Rash spares nasolabial folds

Clinical Features

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Clinical Features

- Fatigue and low-grade joint pains can be constant and not particularly associated with active inflammatory disease
- Whereas, fever, weight loss and mild lymphadenopathy may occur during flares of disease activity

- Think of SLE whenever someone has a multisystem disorder and raised ESR but CRP normal
- If raised CRP in diagnosed SLE, think of infection, serositis, or arthritis

- Raynaud’s phenomenon may antedate other symptoms by months or years
- Features in favour of secondary Raynaud’s phenomenon
  - Age at onset - Over 25 years
  - Absence of a family history
  - Males

- Joint erosion is not a feature of SLE
- Joint deformities may arise as the result of tendon damage - Jaccoud’s arthropathy

- Libman–Sacks endocarditis may occur in SLE
- It is a not due to infection
- Accumulation of sterile fibrin containing vegetations on the heart valves

- Sulfonamides or the oral contraceptive pill may worsen idiopathic SLE

- More than 95% patients have positive Anti-nuclear antibodies (ANA)
- High Sensitivity

- High anti-double-stranded DNA (dsDNA) antibody titre is highly specific
- Positive in ~60% of cases only

- Antibodies against Extractable Nuclear Antigens (ENA) (anti-sm, anti-Ro, anti-La, anti-RNP) - Positive in 20 – 30%

- SLE may be associated with Antiphospholipid antibody syndrome
- Test anticardiolipin antibodies and Lupus anticoagulant in appropriate cases

Disease Monitoring

- 3 tests used to monitor disease activity
    1. Anti-dsDNA antibody titres
    2. Complement C3 & C4 levels
    3. ESR

- Low C3, low C4 denotes consumption of complement
- Hence low C3 and C4, and raised C3d and C4d (their degradation products)

- C3 & C4 can be low in inherited complement deficiency
- Studies of other family members can help to differentiate inherited deficiency *from* complement consumption

- Anti-double-stranded DNA (dsDNA) and anti-Smith antibodies
  - High specificity for SLE
  - Predictor of nephritis and hemolytic anemia
  - Correlates with disease activity

- **False-positive** Venereal Disease Research Laboratory (VDRL) test - high sensitivity, low specificity

- Drug-induced lupus caused by more than 80 different drugs
  - Isoniazid
  - Hydralazine
  - Procainamide
  - Quinidine
  - Chlorpromazine
  - Minocycline
  - Phenytoin
  - Anti-TNF agents

- It is associated with antihistone antibodies in >95% of cases
- Rarely affect kidneys and CNS
- Skin and lung signs prevail
- The disease remits if the drug is stopped

- Conditions in which ANAs are elevated
  - SLE
  - RA
  - Scleroderma
  - Sjögren syndrome
  - Mixed connective tissue disease (MCTD)
  - Polymyositis & dermatomyositis
  - Drug-induced lupus

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