Juvenille Rheumatiod Arthritis

Objectives

- Define JRA
- Describe the etiology and pathogenesis
- Detail the clinical presentations and diagnostic criteria
- Describe the laboratory findings
- Describe the radiologic features
- Plan the management

Introduction

- Commonest rheumatic disorder in children
- American college of Rheumatology classifies it as a category of diseases, with 3 types of onset:

1. Oligo- arthritis (pauciarticular) dx.
2. Polyarthritis
3. Systemic onset dx

Criteria for Diagnosis

- Age of onset < 16yrs.
- Arthritis (Swelling or effusion) presents as ≥2 of limitation of movements, tenderness, or pain on motion, and increased heat in one or more joints.
- Duration of symptoms >6 weeks
- Exclusion of other forms of juvenile arthritis

Etiology

Unknown, but 2 events are considered necessary:

1. Immunogenetic susceptibility – HLA sub-types show increased risk or may be protective.
2. External(Environmental) triggers;

   - Viruses- parvovirus, rubella, EBV.
   - Host hyper- reactivity to specific self antigens (Type 2 collagen),
   - Enhanced T- cell reactivity to bacterial and mycobacterial heat shock proteins.

pathogenesis

- Synovitis characterized by villous hypertrophy, hyperplasia and hyperemia, oedema of sub-synovial tissues.
- Vascular endothelial hyperplasia- with infiltration of mononuclear and plasma cells
- Pannus formation occurs in advanced dx- results in progressive erosion of articular cartilage and contiguous bone.
- T-cell recruitment of different clonal expansions especially HLA DR4, DR8, DR5.
- T- cell activation leads to a cascade of events leading to tissue damage, B-cell activation, complement consumption, cytokine release.

Clinical presentations

- Morning stiffness and gelling
- Easy fatigue –especially early afternoon
- Joint pains – latter in the day
- Joint swelling – warm, restricted motion, may be painful
- Oligo arthritis; Common in lower limbs, but hip unlikely.

- commoner in females, younger age

- Polyarthritis;

  - large and small joints affected (Min 5 joints, max 40) - Assos rheumatoid nodules on extensor surfaces of elbows, and over achilles tendon associated with severe dx.
  - commoner in females, older age
  - Micrognathia – reflect TMJ involvement
  - Cervical spine involvement common, associated with atlantoaxial sub-laxation.

Systemic onset dx

- Quotidian fever, with daily temp spikes of 39C. for ≥ 2 weeks.
- Associated faint erythematous macular rash.
  Associated linear or circular lesions over trunk and proximal extremities.
- Prominent visceral involvement with;
  hepatospleenomegally,
  lymphadenopathy,
  serositis- pericardial effusion
- Arthritis may be pauci/polyarticular

Laboratory findings

Non specific;

- ↑ ESR, ↑ CRP ,
- leucocytosis, thrombocytosis, anaemia of chronic illness.
- + ANA (40-80% poly/ pauci dx, rare in systemic onset dx). Assos with uveitis.
- + Rheumatoid factor; common in older children, polyarticular dx, poor prognosis.
- Abnormal bone mineral metabolism, especially appendicular cortical bone,

  -↑IL- 6 reduces bone formation leading to low osteocalcinin and reduced bone-specific alkaline phosphatase

Radiologic changes

- Soft tissue swellings
- Osteporosis
- Periostitis
- Accelerate regional epiphyseal closure
- Subchondral erosions
- Narrowing of cartilage space
- Bone destruction and fusion
- Severe dx seen in neural arches of C2, C3.
- MRI scan useful.

Complications

- Eye: chronic iridocyclitis, – present with small irregular pupils.
  - Uveitis,
  - Keratopathy
  - Cataracts
- Anaemia- seen in active dx or prolonged dx.
  - Usually unresposive to iron.
  - May be exacerbated by GIT bleed assos with NSAIDS.
  - May be a haemolytic anaemia
  - may be complication of macrophage activation syndrome- assoc with leucopemia, thrombocytopenia, lymphadenopathy, and hepatospleenomegally.
- May develop features of SLE- esp polyarticular dx
- Orthopaedic complications
- Psychosocial issues

Management

Start with the mildest drugs and escalate in severe dx;

- NSAIDS
- Hydroxychloroquine
- Methotrexate
- Immune suppressants
- Steroids- only for severe dx