Approach to a case of arthritis
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objectives

• By the end of this lecture the student should be able to:
  • 1 – Illustrate the differential diagnosis of arthritis
  • 2 – Demonstrate the criteria of JRA
  • 3 – Determine the etiology and the pathogenesis
  • 4 – Present the various clinical types of JRA
  • 5 – Manage a case of JRA
• A 7-year-old boy referred to pedia clinic with fever, inability to walk and Rt knee swelling. The mother gives a history of trauma while he was playing football 5 days ago.
• A 10–years old female referred to the emergency room with pallor, skin rash, fever and joint pain with limitation of movement of both knees, elbows, and ankles joints for two weeks. The child was well and good before this date.
Case:
a 10-year-old boy had bloody diarrhea month ago and for the last 2 weeks is complaining of swelling of his left and right wrists, his left ankle, and left and right 3rd. He has been a febrile and without rash.
Polyarticular

Migratory arthritis

Yes

Fulfills Jones criteria for ARF

YES

Acute rheumatic fever

No

Viral associated arthritis
Lyme disease

Yes

Systemic JRA
Systemic lupus erythematosus
Viral associated arthritis
Leukaemia
Vasculitis (polyarteritis nodosa)

No

Obtain ophthalmology and rheumatology consults
Polyarticular JRA

No

CBC with differential
ESR
ANA
RF

Systemic symptoms present
juvenile rheumatoid arthritis
Case: One swollen joint

A three year old girl presents with two-month history of swollen left knee

- No history of fevers
- No rash
- No weight loss
- No other joints are involved
- CBC and differential within range
- ESR, CRP, Ig within range
- LDH within range
- ANA titer 1:320
- RF negative
Case: Many swollen joints

Consider the case of a 6 year old girl who presents with multiple swollen joints, morning stiffness and mild anemia.

You must think of Polyarticular JRA.
ACR criteria

• Age at onset <16 year
• Arthritis in one or more joints
• Duration 6wks or longer
• Onset type defined by type of disease in first 6 months
  – Polyarthritis: 5 or more inflamed joints
  – Oligoarthritis: <5 inflamed joints
  – Systemic: arthritis with characteristic fever
• Exclusion of other forms of juvenile arthritis
Etiology

• Unknown

• Two events are necessary
  – Immunogenetic susceptibility
  – Environmental trigger
Pathogenesis

– Believed that RA is an autoimmune disease triggered by exposure of a genetically susceptible host to an unknown arthritogenic antigen.

Autoimmune reaction

Activation of CD4+ T cells and lymphocytes and the release of inflammatory mediators and cytokines

Destroy the joints
Rheumatoid arthritis

Pannus formation
Clinical features

- Morning stiffness, easy fatigability particularly after school, joint pain later in the day, joint swelling, warm, lacks of full range of motion.
- Large joints are affected (knees, ankles, elbows and wrists)

**Three major types** of onset are described according to the presentation during the first 6 months of the disease,

- Pauciarticular (four or fewer joints involved)
- Polyarticular (5 or more joints involved)
- Systemic (with fever and rash)
Pauciarticular

- 4 or less joints (usually large) involved in 1st 6 months of disease in asymmetrical fashion.
- Commonest type 60% of affected children
- Joint swelling is common than joint pain

**Two subtypes :**

**Type I (common)**
- Age : 3 - 5 years (girls)
- Commonly affected sites : knees, ankle and elbow
- Iridocyclitis (25%)

**Type II**
- Age : older than 8 years
- Commonly affected sites : large joints of lower extremities
Note: significant suprapatellar swelling (effusion)
Polyarticular

- Occurs in 30% of patients
- Common in girls
- $\geq 5/\text{%}$ both large and small joints involved within the first 6 months of onset
- Joint pain is common than joint swelling

**Two subtypes:**

**Rheumatoid factor positive:**
- Onset: late childhood or early adolescence
- Joints involved: small joints of the hand (MCP and first interphalangeal joints)
- Presence of rheumatoid nodules
- Uveitis (5%)

**Rheumatoid factor negative:**
- Onset: at any age
- Joints involved: knees, wrists and hip
- Rheumatoid nodules absent
Systemic onset

- 10% of affected children
- Younger age: median age 4 years
- Systemic features precede weeks or months before articular manifestations
- Prominent visceral involvement
- **Characterized by** intermittent fever with daily peak of high temp (40 °), with a characteristic twice daily peak for minimum of 2 weeks
• Each febrile episode is accompanied by faint erythematous maculopapular rash (2-5mm) with central clearing and more prominent over trunk (common) and proximal extremities (salmon-colored).

• Koebner phenomena - cutaneous hypersensitivity to superficial trauma.
Investigations

• Blood
  – Elevated WBC and platelet count
  – normocytic normochromic
    • Both +ve in acute and systemic onset
  CRP/ESR
    • Useful for following therapeutic efficacy,
    • Indicator of inflammatory response
  – Elevated ANA
    • In 40-85% , unusual with systemic onset
    • Associated with increased risk of chronic uveitis
Radiology:

- Show soft tissue swelling,
- periarticular osteopenia,
- loss of joint space,
- erosions
- Deformity
- Radial deviation of the wrist
- Ulnar deviation of the fingers
- Swan neck deformity (flexion – hyperextension deformity).
Complications?
Management

• Goals
  – Decrease chronic joint pain and inflammatory process
  – Establish the child in a pattern of adaptation that is as normal as possible
  – Minimal adverse effect of drugs
  – Normal growth and development
• **Principles**
  – It is a multidisciplinary approach involving different specialities—physiotherapist, occupational therapist, orthotists, nurses, psychologist, social worker, orthopedic surgeon, GP, pain management team
  – Treatment is prolonged for many years
  – Begin with simplest, safest and most conservative methods. If this is inadequate choose other agents in orderly fashion
Management of juvenile Rheumatoid Arthritis

• Early aggressive suppression of inflammation is the principle underlying treatment

• Recent changes
  – Increased use of intra articular and IV steroid
  – Establishment of methotrexate as first line
  – Biological and newer treatment for resistant diseases
Cont..

• NSAID
  – Mainstay
  – COX 1 inhibitors- naproxen, ibuprofen, indomethacin, diclofenac piroxicam
  – COX 2 inhibitors- rofecoxib, valdecoxb: have less GI symptoms: not been fully evaluated in children
  – Treatment must continue for at least 2-3 mo before decision to switch over to other NSAID
• Methotrexate
  – Highest response rate (60-80%), less with systemic onset
  – 15-20mg/m²/week

  – Adverse effects: 13% have GI symptoms, liver toxicity (serum transaminase and albumin every 4-8 wks), nausea, vomiting, mouth ulcers, loss of appetite, alopecia, leucopenia
GLUCOCORTICOIDS.

• Prednisolone 1-2mg/kg: severe unremitting arthritis, systemic manifestations (pericarditis, myocarditis,), and rapidly progressive disease
• Often combined with NSAID or others
• Ophthalmic steroid for treatment of chronic uveitis.
Other aspects

• Ophthalmic check up
  – Girls <6 years with ANA positive pauciarticular arthritis screened for uveitis at 3-6 months interval
  – Other forms once at the time of diagnosis and annually thereafter

• Dietary evaluation and counseling to ensure adequate calcium intake

• Physical and occupational therapy