Rheumatic disease

Learning objectives

- To have some knowledge about the presentation of some of rheumatological problem like juvenile rheumatoid arthritis and HSP and how they differ from Rheumatic fever
- How to diagnose and treat JRA HSP and rheumatic fever
- Some information about the drug used to treat JRA, HSP and rheumatic fever
- How to approach to a child presented with arthritis
- To council a family who have JRA or Rheumatic fever

Rheumatic diseases are defined by the constellation of results of the physical examination, autoimmune marker and other serologic tests, tissue pathology, and imaging

SYMPTOMS SUGGESTIVE OF RHEUMATIC DISEASE

- Fever : Systemic JIA, SLE, vasculitis, acute rheumatic fever
- Arthralgia : JIA, SLE, rheumatic fever, JDM, vasculitis
- Weakness : Juvenile idiopathic arthritis, SLE
- Chest pain : JIA
- Fatigue : JIA
- Back pain

Signs Suggestive of Rheumatic Disease

- Malar rash : SLE,JDM

- Oral ulcer : SLE,Behcet disease
- Purpuric rash : vasculitis ,HSP
- Gottron papule : JDM
- Arthritis : JIA, SLE, Rheumatic fever

Diagnostic testing

- CBC
- Autoantibodies tests
- Imaging

Pharmacological treatment

- Nonsteroidal Antiinflammatory Drugs
  Celecoxib, Meloxicam
- Nonbiologic Disease-Modifying Antirheumatic Drugs
  Methotrexate , Leflunomide, Glucocorticoids
- Biologic Agents
  TNF antagonists :Etanercept, adalimumab
- Modulator of T-Cell Activation
  Abatacept

- B-Cell Depletion : Rituximab
- Cytotoxics : Cyclophosphamide

Juvenile Idiopathic Arthritis

- Juvenile idiopathic arthritis (JIA) is the most common rheumatic
- disease in children and one of the more common chronic illnesses of Childhood

- The worldwide incidence of JIA ranges from 0.8-22.6/100,000 children per year

classification

	Percentage	Sex predominace	Age of onset
Oligoarthritis	40-50%	Gils >boys	2-4 years
Polyarthritis	25-30%	Gils >boys	Bimodal 2-4 years 10-14 years
Systemic JIA	5-15%	No sex predominance	1-5 years

ETIOLOGY

The etiology and pathogenesis of JIA are not completely understood, though both immunogenetic susceptibility and an external trigger are considered necessary.

Possible nongenetic triggers include bacterial and viral infections, enhanced immune responses to bacterial or mycobacterial heat shock proteins, abnormal reproductive hormone levels, and joint trauma.

Clinical feature

Initial symptoms may be subtle or acute and often include

- morning stiffness with a limp
- gelling after inactivity.
- Easy fatigability
- poor sleep quality may be associated

Definition of arthritis

It is either Intraarticular swelling or presence of 2 or more of the following

- limitation in range of motion
- Tenderness
- pain on motion

Criteria for the Classification of Juvenile Rheumatoid Arthritis

Age at onset: <16 yr

Arthritis in ≥1 joint

Duration of disease: ≥6 wk

Onset type defined by type of articular involvement in the 1st 6 mo after onset:

Polyarthritis: ≥5 inflamed joints

Oligoarthritis: ≤4 inflamed joints

Systemic-onset disease: arthritis with rash and a characteristic quotidian fever

Exclusion of other forms of juvenile arthritis

Oligoarthritis

It is subdivided into

- Persistent oligoarticular JIA (disease never develop in more than 4 joints
- Extended oligoarticular JIA (disease extended in more than 4 joints after 6 months and it carry bad prognosis

Poly articular JIA

Arthritis involve upper and lower extermities it is subdivided into 2 types

- RF +ve type
- RF –ve type

Case

A 2-year-old boy presented with a high fever and malaise.

A salmon-coloured rash was present at times of fever

Investigation showed markedly raised acute-phase reactants.

Shortly afterwards, he developed severe polyarthritic

joint disease.

What is the diagnosis?

Systemic JIA

Arthritis in ≥1 joint with, or preceded by, fever(quotidian) of at least 2 wk

accompanied by ≥1 of the following:

- Evanescent (nonfixed) erythematous rash
- Generalized lymph node enlargement
- Hepatomegaly or splenomegaly or both
- Serositis

Diagnosis

- JIA is clinical diagnosis but you should send lab test to exclude other diseases

- CBC
- ESR ,CRP.
- Autoantibody titer such as ANA
- RF
- Imaging like x-ray and MRI

Complication

- Chronic anterior uveitis
- Flexion contractures of the joints
- Growth failure
- Constitutional problems
- Osteoporosis (Multifactorial aetiology, including diet, reduced weight bearing, systemic corticosteroids and delayed menarche.

Management

There is need for education and support for the child and family, physical therapy to maintain joint function, and links to other specialties including ophthalmology, dentistry and orthopedics.

Medical management

NSAID (4-6 weeks)

Joint injection (steroids)(under ultrasound guidance – effective, first-line treatment for oligoarticular JIA;

Methotrexate (DMARDs)Effective in approximately 70% with polyarthritis, less effective in systemic features of JIA. It

is given as weekly dose (tablet, liquid or injection) and regular blood monitoring is required

Biologic(DMARDs)

Biologic medications that inhibit proinflammatory cytokines, such as TNF-α, IL-1, and IL-6(etanrecept,adalimumab)

Systemic coricosteriods

Prognosis

- 50% of patients with JIA have active disease persisting into early adulthood, often with severe limitations of physical function.
- Children with persistent oligoarticular disease fare well, with a majority achieving disease remission.
- Extended oligoarthritis carry poor prognosis
- Those with oligoarthritis and positive ANA has greater risk for development of chronic uveitis

Predictors of severe and persistent disease

- young age at onset,
- RF seropositivity or rheumatoid nodules,
- the presence of anti–cyclic citrullinated peptide antibodies,
- and large numbers of affected joints.

Rheumatic fever

Is a inflammatory condition that occur as complication of infection with group A streptococci such as pharyngitis or skin infection

The annual incidence of acute rheumatic fever in some developing countries exceeds 50 per 100,000 children

Worldwide, rheumatic heart disease remains the most common form of acquired heart disease in all age groups

PATHOGENESIS

- The cytotoxicity theory suggests that a GAS toxin is involved in the Pathogenesis(GAS produces a number of enzymes that are cytotoxic for mammalian cardiac cells, such as streptolysin O)
- An immune-mediated pathogenesis

There is a latent period between the infection and the development of the disease (2-4)weeks

CLINICAL MANIFESTATIONS AND DIAGNOSIS

MAJOR MANIFESTATIONS

- Carditis
- Polyarthritis
- Erythema marginatum
- Subcutaneous nodules
- Chorea

MINOR MANIFESTATIONS

Clinical features:

- Arthralgia
- Fever

Laboratory features:

Elevated acute phase reactants:

- Erythrocyte sedimentation rate
- C-reactive protein
- Prolonged P-R interval

SUPPORTING EVIDENCE OF ANTECEDENT GROUP A STREPTOCOCCAL INFECTION

- Positive throat culture
- rapid streptococca antigen test
- Elevated or increasing streptococcal antibody titer

Initial attack:

2 major manifestations,

or 1 major and 2 minor manifestations, plus evidence of recent GAS infection.

Recurrent attack:

2 major,

or 1 major and 2 minor,

or 3 minor manifestations(high risk comunities)

plus evidence of recent GAS infection

The 5 Major Criteria

Migratory Polyarthritis (The joint involvement is characteristically migratory in nature; that is, a severely inflamed joint can become normal within 1-3 days without

treatment, even as 1 or more other large joints become involved.

Carditis subclinical carditis (defined as without a murmur of valvulitis but with echocardiographic evidence of valvulitis) or clinical carditis (with a valvulitis murmur) as fulfilling the major criterion of carditis

Chorea Sydenham chorea occurs in approximately 10-15%

Frequently subtle, movement disorder. Emotional lability, incoordination, poor school performance, uncontrollable movements, and facial grimacing, all exacerbated by stress and disappearing with sleep

Erythema Marginatum

is a rare (approximately 1%

It consists of erythematous, macular lesions with pale centers that are not pruritic

It occurs primarily on the trunk and extremities, but not on the face

Subcutaneous Nodules

are a rare (≤1% of patients with acute rheumatic fever) finding and consist of firm nodules approximately 1 cm in diameter along the extensor surfaces of tendons near bony prominences

Differential Diagnosis of Acute Rheumatic Fever

Arthritis

- JIA
- SLE
- Reactive arthritis
- Sickle cell disease
- Lyme disease

CARDITIS

- Viral myocarditis
- Infective endocarditis
- Kawasaki disease

CHOREA

- Huntington chorea
- Wilson disease
- Systemic lupus erythematosus

TREATMENT

Antibiotic Therapy

the patient should receive 10 days of orally administered penicillin or amoxicillin or a single intramuscular injection of benzathine penicillin to ensure eradication of GAS from the upper respiratory tract.

Antiinflammatory Therapy

Patients with typical migratory polyarthritis and those with carditis without cardiomegaly or congestive heart failure should be treated with oral salicylates. The usual dose of aspirin is 50-70 mg/kg/day in 4 divided doses PO for 3-5 days, followed by 50 mg/kg/day in 4 divided doses PO for 3 wk. and half that dose for another 2-4 wk.

Patients with carditis and more than minimal cardiomegaly and/or congestive heart failure should receive corticosteroids.

The usual dose of prednisone is 2 mg/kg/day in 4 divided doses for 2-3 wk. followed by half the dose for 2-3 wk. and then tapering of the dose by 5 mg/24 hr. every 2-3 days.

Sydenham Chorea

Sedatives may be helpful early in the course of chorea;(phenobarbital)

PROGNOSIS

The prognosis depend on

- Clinical presentation on initial episode
- Severity of initial episode
- Presence of recurrence

50-70% of carditis resolve without residual heart defect

the more severe the initial cardiac involvement, the greater the risk is for residual heart disease.

PREVENTION

Primary Prevention

Appropriate antibiotic therapy instituted before the 9th day of symptoms of acute GAS pharyngitis is highly effective in preventing first attacks of acute rheumatic

Secondary Prevention

Secondary prevention requires continuous antibiotic prophylaxis, which should begin as soon as the diagnosis of

acute rheumatic fever has been made and immediately after a full course of antibiotic therapy has been completed

Vasculitis

Childhood vasculitis encompasses a broad spectrum of diseases that share in common inflammation of the blood vessels as the central pathophysiology.

Childhood vasculitis varies from a relatively benign and self-limited disease such as Henoch-Schönlein purpura to catastrophic disease

with end-organ damage as can be seen in granulomatosis with polyangiitis (formerly Wegener granulomatosis

Henoch-Schönlein purpura

Common

- Arterioles and venules, often small arteries and veins
- HSP and the frequency of preceding upper respiratory infections,
- infectious triggers such as group A β-hemolytic streptococcus, Staphylococcus
- aureus, mycoplasma, and adenovirus have been suspected

CLINICAL MANIFESTATIONS

- rash: palpable purpura

skin lesions are usually symmetric and occur in gravity-dependent areas (lower extremities) or on pressure points (buttocks

- Subcutaneous edema localized to the dorsa of hands and feet, periorbital area, lips, scrotum, or scalp is also common.
- Musculoskeletal involvement, including arthritis and arthralgias,
- Gastrointestinal manifestations of HSP occur in up to 80% of children with HSP and include abdominal pain, vomiting, diarrhea, paralytic ileus and melena; and intussusception
- Renal involvement occurs in up to 50% of children with HSP, manifesting as microscopic hematuria, proteinuria, hypertensio
- Neurologic manifestations of HSP, caused by hypertension or central nervous system (CNS) vasculitis, may also

DIAGNOSIS

The diagnosis of HSP is a clinical one

LABORATORY FINDINGS

Common but nonspecific findings include leukocytosis, thrombocytosis, mild anemia, and elevations of erythrocyte sedimentation rate (ESR) and C-reactive protein

TREATMENT

Treatment for mild and self-limited HSP is supportive,

Steroids are most often used to treat significant gastrointestinal involvement or other life-threatening manifestations

intravenous immune globulin and plasma exchange are sometimes used in the setting of severe disease

PROGNOSIS

Overall, the prognosis for childhood HSP is excellent,