ACQUIRED HEART DISEASE

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Acute Rheumatic Fever

Introduction

Acute rheumatic fever (ARF) is an immune-mediated disorder that follows infection of the pharynx by group A β-haemolytic streptococci. It is more common in developing countries and in areas with overcrowded living conditions. It affects the heart, joints, skin and central nervous system. The sequelae occur 10 days to several weeks after the onset of sore throat. Chronic rheumatic valvular heart disease is the major consequence of acute rheumatic carditis.

Etiology

- At risk age group: 5-14 years; highest 10-14 years, followed by 5-9 years
- Equally common in boys and girls

Epidemiology

- Group A β-haemolytic streptococci: pathogenic strains are M type, 1, 3, 5, 6, 18, 19, 21. The pathogenesis of ARF remains incompletely understood. Current evidence supports the view that ARF is the result of an autoimmune response to pharyngeal infection with Group A β-haemolytic streptococci via molecular mimicry theory (streptococcal cell wall protein and carbohydrate share molecular mimicry with human connective tissue of the heart, brain and joints) in genetically predisposed individuals. Among 0.3 – 3% of people with Group A β-haemolytic streptococci pharyngitis develop ARF, depending on genetic predisposition and virulence of infecting strain. Mitral valve is the most common valve affected in ARF, followed by aortic and tricuspid valves. Pulmonary valves are almost never affected in rheumatic heart disease (RHD). Streptococcal skin infection does not lead to ARF.

Risk factors

Risk factors of acquiring ARF include poor socioeconomic status overcrowding, poor nutritional status and unhygienic surroundings and limited access to healthcare. Streptococcal upper respiratory tract infection can also occur in epidemics which usually involves highly virulent strain.

Clinical features and diagnosis

Symptoms occur about 1-5 weeks (average 2-3 weeks) after initial sore throat. History of preceding sore throat is present in 50% of people. Family history of ARF can be positive. Most patients with ARF present with a combination of fever, joint and cardiac involvements. Chorea and erythema marginatum are rare in children.

- Fever: 90% of patients
- Carditis: present in 50-70% of children with RF. Signs of active carditis include:
  - Endocarditis: pansystolic murmur of mitral valve regurgitation or early diastolic murmur of aortic regurgitation: mitral valve is most commonly involved.
  - Degree of cardiac involvement varies in severity, ranges from mild, transient cardiac involvement to fulminant, potentially fatal exudative pericarditis.
  - Due to wide availability of echocardiography, the Revised criteria (2015) recognizes echocardiographic evidence of subclinical carditis (without a murmur of valvulitis but with echocardiographic evidence of valvulitis, diagnosed in additional 12-21%) or clinical carditis (with a murmur) as fulfilling the major criteria of carditis.
  - Migratory polyarthritis: seen in 75% of cases. Joints commonly involved are knees, ankles, elbows and wrists. The affected joints become red, hot and swollen. It is characteristically described as migratory polyarthritis – may resolve spontaneously in one joint within 24 hours only to appear in another. Menaorheorsitis is a minor criteria, the child commonly responds dramatically (within 12-24 hours) to salicylates. The degree of arthritis does not correspond to the severity of cardiac involvement. Prognosis is good.
  - Erythema marginatum: non-itchy macules with serpiginous pink borders seen on the trunk and inner thighs. It is transient and is an early manifestation of ARF. It is difficult to identify in patients with dark skin.
  - Subcutaneous nodules: are firm, non-tender and are commonly seen on the posterior surface of large joints (knees, elbows, wrists, along the spinous processes of the vertebrae); seen in 10-15% of patients with RF, and appear around 3-6 weeks after the onset of ARF. They are almost always associated with carditis.
  - Sydenham chorea is a late finding, occurring 3 months after the onset of illness. It is purposeless, involuntary, bilateral movements that disappear during sleep. The features include facial grimace, protruding tongue (Wormian tongue) irregular contractions of hand muscles on active grip. Emotional lability may be the first feature. Sydenham chorea is usually self-limiting; lasts for 2-6 weeks. However, emotional lability and poor concentration may last for years.
  - Revised Duke’s Jones criteria. No single clinical feature or laboratory test is diagnostic of ARF. The Jones criteria provide a framework to make a diagnosis (Table 35.1).

Recurrence of rheumatic fever is highest in the first few years after an initial attack and is more common in young children. In a patient with established heart disease, it is characterized by:

  - Endocarditis: a change in pre-existing murmur, emergence of a new murmur.
  - Myocarditis: worsening cardiac failure or cardiomegaly.
  - Pericarditis: chest pain, pericardial effusion or pericardial friction rub.

Recurrence: ARF may not fulfill Duckett Jones criteria for diagnosis.

Investigation

- Raised acute phase reactants: ESR, CRP.
- Evidence of preceding streptococcal infection: increased in