Dr. Deepak Suman
M.D (K.C) NIA, Jaipur

RHEUMATIC FEVER :⇒

Def.: ⇒ Rheumatic fever (RF) is a systemic, autoimmune disease associated with group A-β-hemolytic streptococcal infection, in the course of which the patient develops carditis, arthritis, chorea, subcutaneous nodules and erythema marginatum.

⇒ It is acute, recurrent, inflammatory disease, mainly of children (aged 5-15 yr.), typically occuring 1-5 weeks after group A streptococcal infection.

⇒ The chronic stage of RF involves all the layers of the heart (Pancarditis) causing major cardiac sequelae referred to as rheumatic heart disease (RHD).

Incidence : ⇒ The disease appears most commonly in children between the age of 5 to 15 yr. when the streptococcal infection is most frequent and intense.

Pathogenesis : ⇒ Following exudative streptococcal pharyngitis, rheumatic fever occurs after about 2 weeks. The exact pathogenetic pathway by which streptococcus leads to rheumatic inflammation is not known. Various mechanisms proposed are :-

① Direct toxic action by streptococcus or its "L form" lacking the cell wall.

② Allergic reaction to the organism or its Product.

③ Auto immune reaction :- There is similarity between the carbohydrate of the streptococcus and some Proteins of heart valves causing the immune reaction This is the most accepted theory.

Pathology : ⇒ RF can involve any or all the layers of the heart (Pancarditis). Involvement of the valvular endocardium may lead to tiny vegetation on the valves and destruction and fibrosis of valve substance with fusion of tip cusps of commissures leading to stenosis. There is myocarditis and fibrinous Pericarditis (bread-and-butter appearance). Both are self-limiting causing no permanent sequelae.

Microbiology : ⇒ The hallmark of carditis is the "Aschoff" body which is a granulomatous lesion of the endocardium with fibrinoid necrosis. This area is surrounded by Cardiac histiocytes called Aschoff cells or ~~Anti~~ Anitschoff cells.

CLINICAL FEATURES : ⇒

Modified Jones Criteria For Diagnosis of RF.

Major Criteria -

1. Carditis
2. Migratory Polyarthritis
3. Sydenham's Chorea
4. Subcutaneous nodules
5. Erythema nodosum marginatum

Minor Criteria -

Clinical :- Fever
Arthralgias

Laboratory :-
Elevated ESR or CRP
Prolonged PR interval (ECG)

Essential Criteria :-

Evidence of recent group-A Streptococcal infection -

1. Positive throat culture
   or
2. Elevated streptococcal antibody (anti-streptolysin O (ASO), anti-DNAase
   or
3. Rapid antigen detection test
   or
4. Recent Scarlet fever

To Fulfil Jones Criteria -

⇒ 2 MAJOR OR

⇒ 1 MAJOR AND 2 MINOR

PLUS

ANY ONE OF THE ESSENTIAL CRITERIA

Major Criteria : ⇒

(i) Carditis :- Pancarditis, seen in 50-60% of Patients, develops within the first 2 weeks of rheumatic fever. Pericarditis is evidenced by Presence of a Pericardial rub, myocarditis by tachycardia.

(ii) Arthritis (60-75%) ⇒ Flitting and fleeting type of Polyarthritis involving large joints with no residual deformity is seen in 60-75% of Patients & occurs early in rheumatic fever.

(iii) Subcutaneous Nodules : ⇒ Non-tender nodules are seen over bony prominences like elbows, shin, occiput, spine in 3-5% of Patients and occur 3-6 weeks after onset of RF.

(iv) Erythema Marginatum (<5%) : ⇒ Macular lesions with an erythematous rim and central clearing in a bathing suit distribution are seen in <5% of Patients & occur early in RF.

(v) Chorea (Sydenhams chorea) (2-30%) : ⇒ A neurological disorder with rapid, involuntary & Purposeless non-repetitive movements with a self limiting course of 2-6 weeks is more common in females and late manifestation of RF.

Valve involvement in Rheumatic Heart Disease :-

Mitral valve alone - 50%.

Aortic valve alone - 15-20%.

Mitral and Aortic valve together - 35-40%.

Mitral, Aortic and Tricuspid valves - 2-3%.

Pulmonary valve is virtually never involved.

In RHD, Mitral Valve is most commonly involved followed by involvement of Aortic valve.

Investigation & Laboratory Findings : ⇒

→ High ESR
→ Anemia, leucocytosis
→ Elevated C-reactive Protein
→ ASO titre > 200 Todd-units.
→ Anti-DNAse B test
→ Throat culture
→ ECG - Prolonged PR interval, 2° & 3rd degree blocks
ST depression, T inversion.
→ 2D Echo cardiography :- Valve edema, mitral regurgitation, LA & LV dilation

TREATMENT : ⇒

(a) Tab. Aspirin 75-100 mg / kg / day in 4-5 divided doses, till the activity of the disease subsides (ESR becomes normal).

(b) Steroids in dose of 1-2 mg / kg / day if symptoms of RF and / or carditis persist despite adequate aspirin therapy.

(c) Continuous Prophylaxis against recurrent RF with inj.- benzathine Penicillin 1.2 million unit IM every 3-4 weeks.
In patients allergic to Penicillin, tab. Sulfadiazine 1gm daily or tab. erythromycin 250mg twice daily may be given.

⇒ Prophylaxis must continue, up to the age of 25 yr. or 5 yr. after the last attack.

Prevention of Rheumatic Fever :-

Primary Prevention -

→ Benzathine Penicillin- Once ~~daily~~ only
< 27 kg - 6,00,000 Units
> 27 kg - 12,00,000 Units

→ Penicillin - V
Children - 250mg tid for 10 day
Adolescents / Adults - 500mg tid for 10 days

Secondary Prevention :-

→ Benzathine Penicillin - 12,00,000 Units IM - Every 3/4 weeks
→ Penicillin-V- 250mg bid daily
→ Sulphadiazine - < 27 kg - 0.5g : > 27 kg - 1 gm once daily.

"STAY HOME & STAY SAFE"