Rheumatic Fever

Rheumatic fever is an immunologically mediated inflammatory disease that typically develops two to three weeks after an episode of group A streptococcal pharyngitis due to antibodies directed against the M proteins of streptococci that cross react with self antigens in the heart, joints, skin, and brain. In rheumatic heart disease, distinctive lesions occur in the heart called Aschoff bodies which are foci of primarily T lymphocytes, occasional plasma cells, and enlarged activated macrophages called Anitschkow cells. The cross reactivity of the antibodies can cause myocarditis, which can cause cardiac dilation and can evolve to functional mitral valve insufficiency or fulminant heart failure. Myocarditis is typically the cause of early death due to rheumatic fever. Valvular damage is also common, especially in chronic rheumatic fever. Rheumatic fever can be diagnosed based on the revised Jones criteria and can be made when two of the major criteria, or one of the major criterion plus two minor criteria are present when there is evidence of streptococcal infections. Major criteria include migratory polyarthritis, carditis, subcutaneous nodules, erythema marginatum and Sydenham’s chorea while minor criteria include fever, arthralgia, elevated ESR, leukocytosis, ECG changes (prolonged PR interval), and a known previous episode of rheumatic fever.



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Pathophysiology

Pharyngeal Infection with Strep Pyogenes (Group A Strep)
Pharaoh with Stripper-pie-genie A-apple
Rheumatic fever is an immunologically mediated inflammatory disease that typically develops two to three weeks after an episode of group A streptococcal pharyngitis due to antibodies directed against the M proteins of streptococci that cross react with self antigens in the heart.

Rheumatic Heart Disease
Roman Heart
Rheumatic fever is an immunologically mediated inflammatory disease that typically develops two to three weeks after an episode of group A streptococcal pharyngitis. Acute carditis can occur and may progress over time to chronic rheumatic heart disease, commonly with valvular abnormalities.

Signs and Symptoms

Aschoff Bodies
Ash-pile
In rheumatic heart disease, distinctive lesions occur in the heart called Aschoff bodies which are foci of primarily T lymphocytes, occasional plasma cells, and enlarged activated macrophages called Anitschkow cells. During acute rheumatic fever, Aschoff bodies and diffuse inflammation can be found in any of the layers of the heart.

Anitschkow's Cells
A-knitting-cow
Anitschkow cells are plump activated macrophages that have abundant cytoplasm and central round nuclei that may become multinucleated. They have an ovoid nucleus and chromatin that is condensed toward the center of the nucleus in a wavy rod-like pattern that to some resembles a caterpillar. Anitschkow cells are pathognomonic for rheumatic fever.

Early Death Due to Myocarditis
Gravestone with Mayo-heart-card
Typically, death in acute rheumatic fever is caused by myocarditis, which can cause pericardial friction rubs, weak heart sounds, tachycardia, and arrhythmias. Myocarditis can also cause cardiac dilation and can evolve to a functional mitral valve insufficiency or fulminant heart failure.

Migratory Polyarthritis
Migrating Polly-King-Arthur
Migratory polyarthritis of large joints occurs when one large joint after another becomes swollen and painful for few days and then subsides spontaneously. Migratory polyarthritis is one of the major manifestations of rheumatic fever.

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Subcutaneous Nodules

Sub-q-tips

These are firm but painless nodules that consist of collagen fibers over bones or tendons that can appear on the back of the wrist, outside elbow, and anterior knees. Subcutaneous nodules are one of the major manifestations of rheumatic fever.

Erythema Marginatum

Red Marching-band

Erythema marginatum is a reddish rash that begins as macules on the trunk or arms and spread outward and clear in the middle to form rings. The rings continue to spread and coalesce with other rings but typically spare the face. Erythema marginatum is one of the major criterion of rheumatic fever.

Chorea

Korean-flag

Sydenham's chorea describes a characteristic set of rapid movements of the face and arms that lack purpose. This can occur late in the disease and is a major manifestation of rheumatic fever.

Diagnosis

Elevated ESR

Up-arrow ESR tubes

Erythrocyte sedimentation rate is the rate at which red blood cells sediment in a one hour period and is a nonspecific marker of inflammation. ESR is typically elevated in rheumatic fever and is one of the minor criterion.

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