

Acute Rheumatic Fever (ARF); Let's Think Beyond Old Believes! A Simple Reasoning Behind a Huge Fact

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ABSTRACT

Acute rheumatic fever (ARF) is historically defined as an abnormal immunologic reaction secondary to streptococcal infection. Aschoff's bodies were suggested as pathognomonic findings in endomyocardial biopsies in 1904 by Ludwig Aschoff. Medical terminology of rheumatic heart diseases widely used interchangeably for any valvular or myocardial diseases for decades attributed to the kingdom of streptococcus associated heart attacks as the sole rheumatic diseases centuries ago. When the number of all rheumatic conditions were less than two or three. Considering to the friable connective tissue of valvular leaflets, this fact may make heart valves vulnerable in rather non-ARF rheumatic diseases. Before discovering conditions with potential valvular involvement during last 60 years, possibly most of them were ascribed to ARF-induced rheumatic heart disease and being managed accordingly. Critical issue in ascribing a diagnostic tool (Aschoff's body) as a pathognomonic is domination on all mimicking conditions and full clinical expertise on all aspects of updated medicine. We think that most cases of so-called ARF may actually be diseases other than ARF and we should be careful in labeling patients with joint or valvular heart diseases as ARF. Remarkable decrease in incidence of ARF itself may be in part due to training and worldwide distribution of modern rheumatologist with more precise diagnoses.

Key words: Rheumatic fever; Rheumatic heart diseases; Challenges in diagnosis; Connective tissue diseases

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EDITORIAL

More than 100 years have elapsed after using terminology of rheumatic fever and according to some notions, it seems that a huge amount of changes happened in epidemiology of this basically infectious disease during last decades. An important issue is remarkable decrease in incidence of ARF despite lack of effective vaccination for this potential infectious diseases^[1] Previous reports indicates pathognomonic findings for this heart-joint attacking disease. Aschoff bodies were suggested as pathognomonic findings in endomyocardial biopsies around year in 1904 by Ludwig Aschoff. These micronodules are consisting granulomatous reaction with lymphocytic and occasional plasma cell infiltration and characteristically abnormal macrophages surrounding necrotic center.

Medical terminology of rheumatic heart diseases started and widely used interchangeably for any valvular or myocardial diseases for decades attributed to the kingdom of streptococcus associated heart attacks as the unique rheumatic diseases centuries ago. When the number of all rheumatic conditions were less than two or three. I mean gout, ARF and possibly syphilitic arthritis^[2] That times and shortly after that, campy issue of differentiation between two major and different concepts of joint conditions (rheumatoid arthritis as prototype of inflammatory joint disease and osteoarthritis as prototype of degenerative ones) was challenging. By the way, some researchers believe that pathologic findings of Aschoff bodies could be similar as in nodules elsewhere in the body^[3] No published article we found regarding pathognomonic findings in subcutaneous nodules in the setting of rheumatic disorders. Moreover, this so-

called pathognomonic feature of ARF is never ever clinically applicable unless after post-mortem period.

Considering to friable connective tissue of valvular leaflets, this fact may make heart valves vulnerable in rather all chronic (non-ARF) rheumatic disease spectrum^[4] Before discovering conditions with potential valvular involvement, possibly most of them were ascribed to rheumatic heart diseases and being managed accordingly^[5,6].

Another confounding factor for establishing ARF as clinical diagnosis of joint problems was considerable response of some joint conditions to parenteral penicillin. This in part may be due to anti-inflammatory activities of PCN according our reports^[7].

It seems that a critical issue in ascribing a diagnostic tool as a pathognomonic is domination on all mimicking conditions and full clinical expertise on all aspects of updated medicine. This is the case when in modern rheumatology era, we are challenging with more than 100 different rheumatic conditions mostly introduced or developed after 1950s and major advances in the field of rheumatology emerged after years 1950s^[8] The role of unbelievable development of laboratory or immunologic advances during last decades in differentiating different kinds and subtypes of rheumatic categories could not be underemphasized.

This is interesting while ARF is an acute and clinical disease after a streptococcal infection with known clinical diagnostic criteria, Aschoff bodies are claimed to be pathognomonic for ARF in a case of fever of unknown origin (FUO)^[9] While according to most experiences by experts undifferentiated connective tissue diseases are more eligible to be a candidate for FUO^[10,11].

For more clarification of hidden aspects of this topic we are conducting two projects; effects of crystalline PCN in immune effector cells and the other comprehensive review of patients with diagnosis of ARF in a cohort study. In the last study (under writing) we are showing that near all cases labeled as ARF during last 30 years, none of them had convincing evidence for ARF. We showed that major number of them has some classic or undifferentiated connective tissue diseases. (Unpublished data)

Taking together we can conclude that we think that most cases of atypical ARF may actually be diseases other than ARF and we should be careful in labeling patients with joint or valvular heart diseases as ARF. Remarkable decrease in incidence of ARF may be in part due to educating and worldwide distribution of modern

rheumatologist with more precise diagnoses. This challenging issue basically recalls The Emperor's New Clothes story!

CONFLICT OF INTERESTS

There are no conflicts of interest with regard to the present study.

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