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Review

Diagnostic criteria of acute rheumatic fever

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ABSTRACT

Acute rheumatic fever is an inflammatory sequela of Group A Streptococcal pharyngitis that affects multiple organ systems. The incidence of acute rheumatic fever has been declining even before the use of antibiotics became widespread, however the disease remains a significant cause of morbidity and mortality in children, particularly in developing countries and has been estimated to affect 19 per 100,000 children worldwide. Acute rheumatic fever is a clinical diagnosis, and therefore subject to the judgment of the clinician. Because of the variable presentation, the Jones criteria were first developed in 1944 to aid clinicians in the diagnosis of acute rheumatic fever. The Jones criteria have been modified throughout the years, most recently in 1992 to aid clinicians in the diagnosis of initial attacks of acute rheumatic fever and to minimize overdiagnosis of the disease. Diagnosis of acute rheumatic fever is based on the presence of documented preceding Group A Streptococcal infection, in addition to the presence of two major manifestations or one major and two minor manifestations of the Jones criteria. Without documentation of antecedent Group A Streptococcal infection, the diagnosis is much less likely except in a few rare scenarios. Carditis, polyarthritis and Sydenham's chorea are the most common major manifestations of acute rheumatic fever. However, despite the predominance of these major manifestations of acute rheumatic fever, there can be significant overlap with other disorders such as Lyme disease, serum sickness, drug reactions, and post-Streptococcal reactive arthritis. This overlap between disease processes has led to continued investigation of the pathophysiology as well as development of new biomarkers and laboratory studies to aid in the diagnosis of acute rheumatic fever and distinction from other disease processes.

Contents

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1. Introduction

Acute rheumatic fever (ARF) is a delayed, inflammatory sequela of pharyngitis secondary to Group A Streptococcus infection. The onset of ARF usually occurs approximately two to three weeks following the initial pharyngitis, although in some cases it can present months later. The disease can manifest itself in a variety of presentations including carditis, arthritis, chorea, subcutaneous nodules and a distinctive rash known as erythema marginatum.

ARF was at one time the leading cause of death of children in some parts of the world [1]. Fortunately the severity of the disease was declining even before the availability of antibiotics [2]. Improved medical care and the use of antibiotics have clearly impacted the prevalence of ARF in the developed world. However, the decline in ARF pre-dated the antibiotic era, thus calling into question changes in the virulence of the responsible Streptococcus, presence or absence of certain rheumatogenic types of Streptococcus and possibly the disappearance of a cofactor that may operate in conjunction with the Streptococcus resulting in ARF [2]. Despite the decline documented in the developed world, rheumatic fever remains a serious threat to the health of young people in the developing world.

2. Epidemiology of acute rheumatic fever

Reviews of population based data have estimated that approximately 336,000 cases of acute rheumatic fever occur yearly in children aged 5–14 years, and more than 471,000 cases of ARF occur in all ages [3]. Additionally, it is estimated that 15% of school-age children in developed countries will develop a symptomatic case of Group A Streptococcal pharyngitis annually, whereas the incidence of Group A Streptococcal pharyngitis in less developed countries may be five to ten times that number [3]. The populations affected by Group A Streptococcal disease most frequently are found in developing countries, most commonly in settings marked by poverty or which are medically underserved. Often, accurate data collection is not available and the incidence of ARF in developing countries is likely underreported.

The mean incidence of ARF in the world is 19 per 100,000 [4]. In the United States the incidence of ARF is generally lower than that in developing countries, with reports ranging from 2 to 14 per 100,000 [3–5]. The higher estimates are probably due to recent regional outbreaks documented in certain areas of the United States, including Utah in the 1980s where the incidence among children aged 3 to 17 years approached 12 per 100,000 [6,7]. Other outbreaks have occurred in the developed world. The economic cost of ARF in the United States remains significant and has been estimated to cost anywhere from $224 to $539 million dollars per year [8]. Thus despite overall decline in disease incidence in the United States and other developed countries and the availability of antibiotic therapies, ARF remains a serious healthcare burden in children.

3. Clinical features and diagnostic criteria of acute rheumatic fever

The most current guidelines for the diagnosis of acute rheumatic fever were established in 1992 [9] after revision to the original Jones criteria published in 1944 [10,11]. The goal of these updated criteria was to aid clinicians in the diagnosis of initial attacks of rheumatic fever and to minimize overdiagnosis of the disease. These criteria were not intended to definitively diagnose chronic rheumatic heart disease or predict severity of the disease course.

The diagnostic guidelines for rheumatic fever are divided into major and minor criteria (Table 1). Significantly, the presence of documented preceding Group A Streptococcal infection, in addition to the presence of two major manifestations or one major and two minor manifestations strongly support a diagnosis of acute rheumatic fever. Without documentation of antecedent Group A Streptococcal infection, the diagnosis is much less likely except in a few rare scenarios which will be addressed later in this review.

4. Major clinical manifestations of acute rheumatic fever

4.1. Carditis

Carditis is the most serious manifestation of ARF and ranges from minimal to life threatening heart failure. The carditis that is seen in acute rheumatic fever is almost always associated with a murmur suggesting the presence of a valvulitis [9]. In fact, the absence of a murmur makes the diagnosis of carditis secondary to acute rheumatic fever much less likely. It is important to remember the cardiac manifestations of rheumatic fever may involve the endocardium, myocardium or pericardium, and thus the presentation of rheumatic heart disease is highly variable.

The valvulitis of rheumatic heart disease has several different presentations. Most often it presents as a new apical systolic murmur of mitral regurgitation or a basal diastolic murmur of aortic regurgitation [9]. Presence of an apical mid-diastolic murmur may be seen in association with an apical systolic murmur.

Myocarditis manifested by one of the aforementioned murmurs is consistent with a diagnosis of carditis of rheumatic origin [9]. Myocarditis without valvulitis is typically not secondary to rheumatic fever. Significantly, rheumatic myocarditis may present as congestive heart failure. This severe presentation is a result of left ventricular volume overload secondary to severe mitral or aortic regurgitation. In this case, cardiomegaly may be detected on chest radiography and/or echocardiogram.

Pericarditis may also be seen in acute rheumatic fever, and can be detected on physical exam with auscultation of distant heart sounds or a friction rub. This could complicate auscultation of the murmur of

Table 1
Jones criteria (modified) for the diagnosis of acute rheumatic fever.

Major Manifestations
Carditis
Polyarthritis
Chorea
Erythema marginatum
Subcutaneous nodules
Minor manifestations
Fever
Arthralgias
Elevated acute phase reactants (CRP, ESR)
Prolonged PR interval
Supporting evidence of Group A Streptococcal infection
Positive throat culture
Positive Rapid Streptococcal antigen test
Elevated or rising Streptococcal antibody titer