A Case Report of Acute Cardiac Failure in a Child: An Atypical Presentation

Acute Rheumatic Fever; A Challenging Diagnosis

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Abstract

Acute rheumatic fever (ARF) is recognized as a diagnostic challenge and can present with a wide variety of symptoms. The Revised Jones criteria (RJC) (2015) aids the diagnostic process by identifying key major and minor features (Table 1), however non-conforming cases continue to arise. Consequent misdiagnosis results in over and under recognition of ARF and risks inappropriate management. A significant complication of ARF is rheumatic heart disease; a cause of significant morbidity worldwide. An awareness of unusual cases aids clinical judgement and can help make difficult diagnoses.

We present the case of a nine-year-old Malaysian boy who presented acutely in congestive cardiac failure. He had features consistent with the RJC including carditis, increasing erythrocyte sedimentation rate (ESR), elevated anti-streptolysin O titer (ASOT) and electrocardiogram (ECG) changes. Despite this, he did not fulfil the RJC as ECG findings are non-diagnostic in the presence of carditis. In addition, a raised ASOT lacks significance in areas with a high prevalence of streptococcal infection if the RJC are not met such as in this case. Consequently, due to the atypical nature of the presentation and isolated cardiac features, an alternative diagnosis of rheumatic heart disease was considered. We discuss the diagnostic challenges of ARF and RHD faced in this case.

Keywords

Acute Rheumatic Fever; Rheumatic Heart Disease; Heart Failure; Carditis; Revised Jones Criteria

Background

Acute rheumatic fever (ARF) is an autoimmune response which occurs secondary to a pharyngeal infection with Group A Streptococcus (GAS) [1-5], classically streptococcus pyogenes [6-8]. This response typically occurs within two to four weeks of the primary GAS infection and predominantly affects 5-14 year olds [9, 10]. There are ranges of presenting features which are outlined in the Revised Jones Criteria (RJC). Two major features or one major and two minor features, with evidence of preceding streptococcal pharyngitis, is diagnostic [1]. The severity of symptoms can differ significantly, with some requiring hospitalization, and others remaining asymptomatic. Due to the multi-systemic nature of the condition and diversity in presentation, high clinical suspicion is required in atypical cases to avoid misdiagnosis. Regardless of symptoms or severity, all those diagnosed with suspected ARF should receive treatment to reduce the risk of disease progression and cardiac sequel [1, 11].
The most significant complication of ARF is the development of rheumatic heart disease (RHD). RHD is a multi-valvular cardiac condition which typically occurs 10-20 years after the initial ARF episode [12]. However, the onset of RHD can be accelerated in severe or recurrent cases of ARF [9].

The diagnosis of ARF and RHD can be challenging as there is a significant burden of disease which remains asymptomatic [3, 13]. Echocardiography is the therefore the predominant mode for determining clinical and subclinical cardiac involvement of RHD and ARF [1]. Consequently, screening programs which involve the use of echocardiography are being established [14, 15].

The incidence of ARF is declining in developed countries due to improved infection control measures leading to a reduction in the spread of GAS [16]. Despite this, some areas, such as the Middle East [17] and Malaysia [18], remain affected and are considered moderate-high risk populations [19].

Without the use of secondary prophylaxis ARF can progress to RHD [14]. There continues to be a significant level of global morbidity and mortality associated with RHD, 282,000 new diagnoses and 233,000 deaths worldwide [3]. Understandably, it is essential to have an awareness of both typical and atypical presentations of ARF to allow accurate diagnosis, ensure better disease control and limit progression.

Case Presentation

History and Examination

A nine-year-old, previously well, Malaysian boy presented with a first episode of breathlessness. His symptoms developed rapidly over a four-day period prior to presentation. Initially, the breathlessness was exertional, but progressed until symptoms were present at rest. This was compounded by the onset of intermittent palpitations, orthopnea and paroxysmal nocturnal dyspnea (PND). He had remained a pyrexia with no history of chest pain, syncope and without joint or skin involvement.

Two weeks prior to presentation he had a self-resolving fever and gastroenteritis with no apparent complications. Otherwise, he had no other significant medical history except a sibling death from congenital heart disease.

On clinical examination, evidence of cardiac pathology was identified: signs of moderate respiratory distress tachycardia of 130-140 beats per minute and a heaving cardiac apex beat felt over the 6th intercostal space mid-clavicular line. Auscultation demonstrated a gallop rhythm with pan systolic murmur (Levine scale grade 4/6 [20] heard over the apex and radiating to the axilla and para-scapular region. Pulmonary examination identified bilateral basal crepitations’, with otherwise adequate air entry. Peripherally, findings included a raised jugular venous pulsation (JVP), pedal edema, tender hepatomegaly and adequate peripheral pulses in regular rhythm.

Completion of the examination failed to demonstrate evidence of pyrexia, tonsillar enlargement, pharyngitis, rashes, arthralgia or neurological dysfunction.

Investigations

Baseline investigations showed an unremarkable full blood count (FBC) and C-reactive protein (CRP). Erythrocyte sedimentation rate (ESR) was mildly elevated at 43mm/hr, with levels rising to 80mm/hr at 72 hours after admission.

Due to suspected cardiac involvement, more specific investigations were performed: N-Terminal Pro-Brain-type naturetic peptide (NT-Pro-BNP) levels were highly elevated at 1210pg/ml; where a diagnostic level for cardiac failure in children is 598pg/ml [21, 22]. As ARF was suspected an ASOT was performed, and gave a result of 1250 IU/ml (upper limit normal range between 200-400 IU/ml) [4, 23].

A 12 lead ECG exhibited sinus tachycardia with a P-R interval of 140msec, with no alteration to S-T, T wave or Q-T segments. Unsurprisingly, the chest X-ray supported a diagnosis of cardiac failure due to bilateral alveolar shadowing and pulmonary venous engorgement, although no cardiomegaly was seen.

Echocardiography identified bilateral dilated atria and a dilated left ventricle with good contractility. Study of the vasculature was normal; however there was substantial valvular pathology. The scan identified defective cooptation and thickening of the anterior and posterior mitral valve leaflets which had resulted in severe mitral regurgitation. In addition, mild to moderate aortic incompetence was seen. The scan confirmed moderate tricuspid regurgitation with valvular gradient of 28mmHg. This supports the clinical findings of the pan systolic murmur, raised JVP and hepatomegaly.

Treatment and Follow Up

With ARF as the most likely diagnosis, treatment with oral prednisolone, penicillin and diuretics were...
initiated. He remained closely monitored as a hospital inpatient and showed improvement in symptoms within six days. At discharge, he continued to take penicillin and was entered into a structured follow up program with the pediatric and cardiology teams.

**Discussion**

ARF is a complication of streptococcal pharyngitis, diagnosed using the RJC. The recent update to the RJC differentiates diagnostic criteria between low and moderate-high risk populations. Low risk is defined as “ARF incidence < 2 per 100,000 school-aged children per year or an all-age prevalence of RHD of ≤ 1 per 1000 population per year”[1]. Moderate-high risk is everything outside of this definition. Both Malaysia and Qatar are considered moderate-high risk populations[17-19]. Positive diagnosis using the RJC is made if the patient displays features of two major, or one major and two minor features from the criteria, with evidence of preceding streptococcal infection (Table 1).

<table>
<thead>
<tr>
<th>Low Risk Population Criteria</th>
<th>Moderate-High Risk Population Criteria</th>
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<tbody>
<tr>
<td>Major:</td>
<td>Major:</td>
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<tr>
<td>Carditis (clinical and/or subclinical)</td>
<td>Carditis (clinical and/or subclinical)</td>
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<tr>
<td>Polyarthritis</td>
<td>Arthritis (monoarthritis or polyarthritis; polyarthralgia can be considered as a major manifestation if other causes are ruled out)</td>
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<td>Chorea</td>
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<td>Erythema marginatum</td>
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<td>Subcutaneous nodules</td>
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<td>Minor:</td>
<td>Minor:</td>
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<tr>
<td>Fever (≥38.5°C [≥101.3°F])</td>
<td>Fever (≥38.0°C [≥100.4°F])</td>
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<tr>
<td>Polyarthralgia</td>
<td>Polyarthralgia</td>
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<td>Elevated inflammatory markers (ESR ≥60/mm hour and/or CRP ≥28.57 nanomols/L [≥3.0 mg/dL])</td>
<td>Elevated inflammatory markers (ESR ≥30/mm hour and/or CRP ≥28.57 nanomols/L [≥3.0 mg/dL])</td>
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<tr>
<td>Prolonged PR interval on electrocardiogram</td>
<td>Prolonged PR interval on electrocardiogram</td>
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Such as in this example, it is recognized that not all cases of ARF conform to the RJC and a diagnosis of atypical ARF can be made in spite of this. Differential diagnoses of congenital heart disease, degenerative disorders and infective endocarditis were ruled out in this case.

There are elements of his presentation which support ARF as the primary diagnosis, but other features lean towards RHD.

Although carditis is a presenting feature in half of ARF cases, more than 90 percent have concurrent pyrexia [1] and 75% have arthritis [8] or additional features at presentation. Isolated cardiac symptoms are therefore unusual.

Prior to presentation he had symptoms of gastroenteritis. There was no evidence of a preceding streptococcal infection, such as pharyngitis. Although vomiting and diarrhoea are seen in up to a quarter of cases of streptococcal bacteremia [24], it is not known to directly cause gastroenteritis. Consequently, a history of gastroenteritis would not usually trigger a clinician to consider ARF as the cause of profound cardiac decompensation.

It would be uncommon without a history of severe or recurrent ARF for a diagnosis of RHD to be made at nine years old, although there are cases of asymptomatic ARF progressing to RHD [9].

The cardiac pathology which is classically associated with ARF is pancarditis; affecting the endocardium, myocardium and occasionally the pericardium [1]. Valvular lesions predominantly affect the mitral valve causing mitral regurgitation [25], but many patients exhibit aortic regurgitation and tricuspid involvement as well [8].

On the other hand, the chronic changes of RHD are typically exclusively valvular changes and usually cause mitral stenosis [26]. However, the REMEDY study recognizes that mitral regurgitation and multi-valvular involvement in under 10 year olds is more common than stenosis alone [26]. Concurrent aortic involvement occurs in 20-30% cases and the tricuspid valve is affected in 15-40% [8].

The echocardiography findings in this child demonstrated multi-valvular involvement with severe mitral regurgitation, aortic incompetence and tricuspid regurgitation. Additionally, he had a trial and ventricular dilatation secondary to severe regurgitation, with adequate myocardial contractility. Conversely, the clinical findings and NT-Pro-BNP suggested cardiac failure and myocardial dysfunction.

The ECG had slightly prolonged PR interval, however this is an insignificant finding when carditis is
present [1].

Such extensive valvular involvement would be unusual in RHD, however a lack of pericarditis and evidence of valvular changes such as thickening and coaptation, may indicate a chronic process. Thus, a diagnosis of acute disease superimposed on underlying RHD is possible.

A further positive feature exhibited was a raised ESR, which subsequently elevated over the following days. Although this was not associated with raised CRP, elevated inflammatory markers favor an acute disease process. It is recorded that RHD can be associated with higher CRP than controls, these generally are lower than during the acute disease phase [27].

In addition to this, there was evidence of a possible preceding streptococcal infection due to a raised anti-streptolysin O titer (ASOT). With an upper limit of normal between 200-400IU/ml, an ASOT of 1250IU/ml is significantly raised [4]. Studies have identified that a sharp increase in ASOT is more commonly associated with ARF than CHD [4], and ASOT levels plateau and decline between 6-12 months after initial streptococcal infection [28].

However, in areas with a high prevalence of ARF, elevated ASOT is not considered significant when the remaining RJC are not met [11]. Consequently, in this example, it is difficult diagnose a streptococcal infection with certainty.

Conclusion

Despite evaluating the evidence for diagnosing ARF and RHD, a concrete diagnosis remains difficult. Cases with isolated features, such as acute cardiac failure, rely on an astute clinician to consider ARF as a differential diagnosis and complete the appropriate work-up investigations. With asymptomatic streptococcal infection, we cannot rely on the history to trigger this differential. The RJC helps aid the diagnosis but must be used alongside a high index of suspicion in order to make appropriate diagnoses. An awareness of unusual presentations and atypical cases is important, especially in high prevalent areas, to avoid misdiagnosis. We propose that where there is a presentation of isolated features, consideration should be made towards ARF as a differential diagnosis.

References


