ACUTE RHUMATISMAL MYOCARDITIS MIMICKING AN ACUTE CORONARY SYNDROME

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Abstract
Acute rheumatic fever (ARF) is an autoimmune disorder usually occurring after group A streptococcal pharyngitis. Myocardial involvement during an ARF is usually associated with valvular and/or pericardial disease and has heterogeneous symptoms among which it can mimic an acute coronary syndrome. The diagnosis is based upon the Jones criteria and its management is both therapeutic and prophylactic. We report here the case of a 25-years-old male patient who consulted for an acute chest pain evoking an acute coronary syndrome. The diagnosis of carditis complicating an ARF was accepted over the presence of multiple criteria and the medical outcome was good under specific medication.

Introduction:
ARF usually occurs 3 weeks to several months after a group A streptococcal pharyngitis. It’s an autoimmune reaction that can affect the heart, brain, skin, and joints [1]. Carditis in ARF is considered to represent pan-carditis involving heart valves, myocardium, and pericardium. While valvulitis is the most common feature of ARF, myocarditis is rare and has heterogeneous symptoms that can mislead the diagnosis [2].

Case Report:
We report here the case of a 25-years-old male patient with no cardiovascular risk factors. His medical history includes recurrent episodes of pharyngitis during childhood. The patient was admitted to the emergency department for intense constrictive chest pain that has been evolving for 5 hours. The interrogation found an episode of pharyngitis during the previous 3 weeks before the current history.

On admission, the axillary temperature was 37.4 °C, blood pressure was 107/64mmHg, and the heart rate was 74 beats/min. The patient reported as well polyarthralgia interesting the knees and ankles but with no sign of arthritis.

At the cardiac auscultation, the rhythm was regular, with aortic regurgitation murmur quoted 3/6, lungs were dry. There was no peripheral sign of heart failure and the rest of the physical examination was normal.

Electrocardiogram (ECG) showed a sinus rhythm with ST elevation in anterior, lateral, and DII leads (Figure 1).

Chest X-rays and computed tomography showed no cardiomegaly, pulmonary congestion, or inflammatory findings.
Echocardiography done the same day showed a moderately reduced ejection fraction (EF) of 50% with anterior and lateral walls hypokinesis. The aortic valve was bicuspid type 0 (Figure 1) with mild aortic regurgitation. Mitral and aortic valves were thickened, and there was a mild pericardial effusion.

Biological features found an accelerated erythrocyte sedimentation rate (ESR) at 100 mm the 1st hour, and C reactive protein (CRP) level at 50 mg/l. Troponin was positive at 38 ug/l and Antistreptolysin O (ASLO) titer of 1600 IU/MI.

Cardiac catheterization showed normal coronary anatomy.

Cardiac MRI (CMR) showed a signal intensity increase in T2-weighted images and late gadolinium enhancement (LGE) of nodular aspect in the anterior, lateral and posterior walls at the pericardial and central myocardial sides (Fig. 3). Very discrete hypokinesia of the left ventricular and EF was at 43%. These findings were enough to fulfill Jones criteria, and ARF was diagnosed.

Symptoms were improved under treatment comprising amoxicillin (3g/day for 10 days) and high dose steroid intake: Prednisone (1 mg/Kg) until normalization of ESR then reduction of 5 mg each week. To prevent further relapses, V Penicillin at 500mg twice a day dose was given up to the age of 40.

On the other hand, male descendants of the patient and brothers were convened for bicuspid screening.

The electrical evolution was marked by a negativation of T waves prior to day 4 (Figure 1B), then a normalization of the ECG at day 10. Control echocardiography after 2 months showed a normal wall motion with an improvement of EF up to 60% with no pericardial effusion.

**Figure 1A:** EKG showing normal sinus rhythm with ST elevations in leads anterior, lateral and DII.

**Figure 1B:** EKG showing normal sinus rhythm with T negatives in leads anterior, lateral.
Figure 2: Echocardiographic images assessing bicuspid aortic valve. A: Transthoracic parasternal short-axis view showing bicuspid aortic valve type 0. B: Transthoracic parasternal long-axis view showing abnormal systolic opening (“doming”) of a bicuspid aortic valve.
Discussion:
Although the incidence of ARF and rheumatic heart disease has decreased thanks to the early availability of antibiotics and improved living conditions, they still prevail in developing nations [3, 4]. Cardiac involvement, called carditis occurs in about 50% of patients with ARF. Inflammation mainly involves the valvular endocardium, sometimes the pericardium and rarely the myocardium [4]. Carditis usually happens 3 weeks after a streptococcal throat infection. Since our patient respects this delay, we can consider the diagnosis of ARF.

While valvulitis is easily revealed by the presence of a heart murmur, a heart failure when the lesions of the endocardium are severe, or fortuitously discovered during an echocardiographic examination, myocarditis has heterogeneous and unspecific cardiac signs and symptoms, depending upon the extent of the inflammation and cardiac dysfunction [5]. It can reveal itself in some cases with chest pain evoking angina as it is the case of our patient. Physical signs include a muffling of heart sounds, an early diastolic gallop, or a global heart failure [6].

As for ECG features, myocarditis has unspecific signs including idiopathic supraventricular or ventricular bradycardia or tachyarrhythmias, PR depression when pericarditis is associated. ST-T segment abnormalities, mimicking an acute coronary syndrome is rarely observed [7] as in our case. T wave inversion generally comes after ST-T segment normalization.

Echocardiography helps to study the morphology and function of the valves and cardiac chambers. The signs of myocarditis are unspecific, it may be normal, or shows segmental wall motion abnormalities or diffuse hypokinesis. Pericardial effusion may be present and suggest associated pericarditis [5]. Echocardiography also helps in follow up by assessing biventricular function during resolution or reactivation of the inflammatory process.

CMR gives morpho-functional information similar to echocardiography but provides additional data about tissue characterization [5]. CMR carries the highest diagnostic accuracy when these three parameters are presently known as the Lake-Louise criteria: 1-the presence of myocardial edema represented by a regional or global myocardial signal intensity increase in T2-weighted edema images; 2- early myocardial hyperemia represented by an increased global myocardial early gadolinium enhancement in gadolinium-enhanced T1-weighted image; 3-the presence of myocardial necrosis or fibrosis represented by at least one focal lesion with nonischemic regional distribution in inversion recovery-prepared gadolinium-enhanced T1-weighted images [5]. In myocarditis late gadolinium enhancement location is typically subepicardial, while it is subendocardial in ischemic heart disease, sometimes located at the inferolateral or septal left ventricular wall segments. As for echocardiography, CMR helps to monitor the evolution of myocardial inflammation.
ESR and CRP serum levels may be increased and are among the minor Jones criteria as in our patient. Cardiac troponins can be elevated in ARF induced myocarditis [5]. In adults, we consider that there is an acute streptococcal infection when the ASLO titer is above 250 UI/ml. But its elevation in a 2 weeks interval is more significant. ASLO was positive with an upward trend in our patient, strengthening the ARF diagnosis.

Throat screening for streptococcus is usually negative. But when it’s positive, outside a period of epidemic and is associated with other features, it consolidates the diagnosis of ARF [6].

**Table 1:** Revised 2015 Jones criteria for acute rheumatic fever.

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<tr>
<th>Major Criteria</th>
<th>Low risk populations</th>
<th>Moderate-high risk populations</th>
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<tbody>
<tr>
<td>Clinical or subclinical carditis</td>
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<tr>
<td>Arthritis</td>
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<td>Polyarthritis</td>
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<td>Chorea</td>
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<td>Erythema marginatum</td>
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<tr>
<td>Subcutaneous nodules</td>
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<tr>
<td>Minor Criteria</td>
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<tr>
<td>Polyarthritis</td>
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<tr>
<td>Fever &gt;38.5°C</td>
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<td>ESR &gt;60 mm in the first hour and/or CRP &gt;6 mg/L</td>
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<td>Prolonged P-R interval, age-corrected (unless carditis is a major criterion)</td>
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**ARF’s diagnosis is based upon the 2015 revised Jones criteria:**
AFR is highly probable when we have two major or one major and two minor criteria. The diagnosis of clinical carditis during ARF is based on the presence of any of the following findings: (1) new onset of organic cardiac murmurs, (2) cardiac enlargement, (3) signs of heart failure, (4) pericardial friction rubs or accumulation of pericardial fluid.

As for all myocarditis standard treatment includes symptomatic management of arrhythmias and heart failure, with emphasis on the importance of resting. In ARF induced myocarditis, high dose steroid therapy should be considered since it is highly sensitive to it. However, the prognosis is extremely poor without treatment [6, 8].

Secondary prophylaxis aims to prevent recurrence by eradicating streptococcus with anti biotherapy and a patient’s education.

The duration of secondary prophylaxis depends on the patient’s age, the date of the last attack and the presence and severity of rheumatic heart disease. It is often maintained until the adult age, sometimes for life for patients with severe rheumatic heart disease or previous valvular surgery [9].

V penicillin remains the reference in terms of antibioprophylaxis. It is given every 3 to 4 weeks while oral penicillin is given every day. In the case of allergy, erythromycin can replace penicillin.
Table 2: International recommendation for secondary prophylaxis of acute rheumatic fever [4].

**Conclusion:**
Acute chest pain in adults can result from acute rheumatic fever and mislead the diagnosis in the presence of signs evoking acute coronary syndrome. A good interrogation and examination in addition to biological and echocardiographic features readdress the diagnostic and guide the treatment. Optimal management of this entity is essentially based upon appropriate antibiotics associated with corticoid therapy if myocarditis included which improves the prognosis and outcome.

**References:**
5. ALP Caforio, G Malipiero, R Marcelongo, and S Iliceto, Myocarditis, 2018 Elsevier Inc.