EMERGENCY CASEBOOK

Acute rheumatic fever mimicking an acute coronary syndrome

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An elevated troponin measurement does not always reflect myocardial ischaemia secondary to obstructive coronary artery disease. Troponin levels can also be elevated in other disease states including pulmonary emboli, myo-pericarditis, acute rheumatic fever, and in the critically ill. Thus, patients presenting with chest pain and electrocardiological and biochemical evidence of myocardial necrosis are not always suffering from an acute coronary syndrome.

A 22 year old Caucasian male was admitted as an emergency to the coronary care unit with a 12 h history of retrosternal chest discomfort. The discomfort was maximal 1 h before admission and was not related to respiration or posture. The patient had no risk factors for coronary artery disease. He was taking oral penicillin for an upper respiratory tract infection and had no other past medical history. On examination he looked unwell, was pyrexial, and had a pulse rate of 105 bpm. There was no cardiac murmur or rub on auscultation. Respiratory and abdominal examination was normal. A 12 lead ECG showed ST elevation in leads II, III, aVF, V₅, and V₆ (fig 1).

An ST elevation myocardial infarction was presumed and urgent coronary angiography was undertaken. The epicardial coronary arteries were angiographically normal. However, the 12 h troponin T (TnT) level was elevated at 0.52 ng/ml (normal TnT <0.01 ng/ml). A transthoracic echocardiogram showed mildly impaired left ventricular systolic function due to inferior wall hypokinesia, but no significant valvular lesion was identified. The diagnosis was revised to acute myo-pericarditis. Other results included CRP 143 and an anti-streptolysin O (ASO) titre of 420 IU (normal <200 IU). A throat swab (taken on admission) only identified respiratory commensals.

The clinical diagnosis using the revised Jones criteria was acute rheumatic fever (ARF; myo-pericarditis, fever, and elevated acute phase reactants, on the background of a recent streptococcal infection). On repeat assessment, there was no history of arthralgia and no evidence of erythema marginatum or subcutaneous nodules. The patient made a good clinical recovery and is now under shared follow up between cardiology and infectious diseases.

DISCUSSION

ARF is characterised by an abnormal autoimmune reaction to a pharyngeal group A streptococcal infection with a resulting systemic inflammatory response. Any part of the body can be affected, and although it can occur at any age, it typically presents in childhood, just before adolescence, with a female predominance. The incidence of ARF varies widely throughout the world. Within Europe and North America the incidence is falling, largely due to improved sanitation and the availability of antibiotics, although isolated resurgences

in North America have been reported. Within the developing world, it remains a common cause of acquired heart disease.

There is no single, gold standard, pathognomonic investigation for ARF. The diagnosis of the initial acute attack is therefore clinically based using the revised Jones criteria (table 1).¹ These criteria are only validated for the initial attack. Our patient had evidence of carditis in addition to fever and elevated acute phase reactants on the background of a history of recent pharyngeal infection and a positive anti-streptococcal antibody test.

In almost 50% of cases of ARF, cardiac involvement will be evident although severe decompensated cardiac failure is rare. The long term sequelae of chronic scarring of the cardiac valves producing valvular stenosis and/or regurgitation are well known, although they are not seen in all patients.

In the acute phase, rheumatic fever can affect any layer of the heart (peri-, myo-, or endocardium). Historically, the carditis has been regarded as a pancarditis with cardiac failure occurring secondary to myocyte injury.¹ However, the current evidence base suggests endocarditis is the predominant factor and that valvular incompetence and not primary myocyte injury predisposes patients to severe cardiac failure. This is supported by studies assessing cardiac troponin measurements in patients with ARF. Whilst troponin levels may be elevated to a minor degree in acute rheumatic carditis, this is of no significance in relation to the development of cardiac failure.^{2,3}

The current diagnostic criteria for ARF are clinically and laboratory based. Echocardiographic studies have reinforced the notion that valvular disease rather than myocardial disease is the primary cardiac abnormality responsible for the development of cardiac failure. Due to the difficulty in accurately differentiating between pathological and physiological minor valvular incompetence, echocardiographic parameters have not been incorporated into the diagnostic criteria for acute rheumatic carditis and its role in the diagnosis of ARF remains controversial.

Table 1 Revised Jones criteria for the initial diagnosis of acute rheumatic fever

Major criteria
Carditis
Polyarthritits
Chorea
Erythema marginatum
Subcutaneous nodules
Minor criteria
Clinical
Arthralgia
Fever
Laboratory
Elevated ESR/CRP
First degree atrioventricular block

CRP, C reactive protein; ESR, erythrocyte sedimentation rate.

Figure 1 12 lead ECG showed ST elevation in leads II, III, aVF, V₅, and V₆.

Optimal treatment of ARF remains unclear, largely because of a lack of randomised controlled trials. Due to the presumed inflammatory nature of the condition, anti-inflammatory agents including corticosteroids are frequently used, although a recent Cochrane review showed no benefit in reducing cardiac complications at 1 year in those patients treated with corticosteroids or immunoglobulins compared with placebo.⁴ Salicylates lead to resolution of fever, arthritis, and arthralgia although their use in carditis is not recommended, and the search continues for an effective treatment of ARF. To prevent recurrences of ARF and carditis, long term antibiotic prophylaxis is recommended, with treatment tailored to the patient's individual risk of recurrence. Our patient made a good clinical recovery and is now under shared follow up between the cardiology and infectious diseases departments. Long term phenoxymethylpenicillin as secondary prophylaxis has been prescribed and serial echocardiography will be performed.

SUMMARY

To summarise:

- An elevated troponin level does not always imply an acute coronary event.
- Troponin levels (troponin T and I) may be elevated in ARF although their prognostic significance is unknown.
- ARF is caused by a group A streptococcal pharyngeal infection. ARF never follows a cutaneous infection.
- The incidence of ARF within industrialised countries remains low although isolated resurgences have been reported.
- The diagnosis of ARF is clinically based using the revised American Heart Association endorsed 1992 Duckett Jones Criteria.
- The role of echocardiography in the diagnosis of ARF remains controversial.

- Valvular incompetence, not myocardial necrosis, is the primary cardiac abnormality responsible for the development of cardiac failure.
- Secondary prophylaxis to prevent recurrences is the most effective approach to control ARF and rheumatic heart disease.

For a diagnosis of ARF, evidence of two major criteria or one major and two minor criteria is required with supporting evidence of streptococcal infection, for example, positive throat culture or rapid antigen test for group A streptococcal or raised or rising streptococcal antibody titre.

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