Rheumatic Fever: Atypical Presentation in an Adult

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SUMMARY: A 38 year old Caucasian lady presented with a history of vague ill health, raised ESR and prolonged P-R interval on ECG. These features became normal within a week. Contrary to the usual presentation, this patient developed fleeting arthritis, one of the major diagnostic features of rheumatic fever, towards the end of her illness. It is important to recognise such variants of rheumatic fever, even in retrospect, for the sake of giving penicillin prophylaxis to prevent cardiac morbidity.

Introduction

The incidence of rheumatic fever (RF) has decreased so much in recent years in the West, that it is extremely rare to find such cases in general medical practice. It usually affects children between 5 and 15 years of age. Cases of RF should fulfil at least one major and two minor of the revised Duckett Jones criteria (Table 1) for the diagnosis to be made, supported by culture or serological evidence of recent streptococcal infection (1).

Table 1
Diagnosis of Acute Rheumatic Fever
by Jones Criteria (revised)

Major Manifestations	Minor Manifestations
Polyarthritis	Arthralgia
Carditis	Fever
Chorea	Elevated ESR or CRP
Erythema marginatum	Previous rheumatic fever or rheumatic heart disease
Subcutaneous nodules	Prolonged P-R interval
Supporting evidence of preceding streptococcal infection	

In most cases enough features are present to make a working diagnosis at the onset of the illness. In some cases, however, it is difficult to reach a definite diagnosis due to the lack of sufficient features in the early stages. A period of careful follow up with a high index of suspicion for evolving features of RF, will help reach a diagnosis in such cases. I have presented here one such case of an adult female where the diagnosis of RF was not possible early in her illness, but who developed migratory joint symptoms later. It emerged on review of the overall clinical picture that she had sufficient diagnostic features to make a firm diagnosis of RF.

Case Report

A 38 year old Caucasian housewife was admitted to the hospital with a history of dizziness on standing, feeling faint and general malaise. She had had a flu-like illness with sore throat a week before the onset of these

symptoms. She had no other relevant past medical history or family history. She denied taking any drugs, apart from aspirins at the beginning of her illness.

On examination she had a variable pulse rate of 80-100 per minute, with postural hypotension (recumbent BP 110/60 and on standing 80/60). Her skin was tanned. Otherwise examination was normal.

Investigations revealed a WBC count of $14.3 \times 10^9/L$, haemoglobin 14.5 gm%, ESR 100mm/1st hour. An ECG showed marked prolongation of P-R interval (0.38 secs) (Fig 1a + b). Other investigations, including urea, electrolytes, plasma cortisol, liver function test, blood glucose, cardiac enzymes, serum calcium and chest X-ray were normal. An echocardiogram did not reveal any evidence of aortitis or vegetation on the valve cusps. The result of her autoantibody profile (ANF, rheumatoid factor and syphilis serology) were reported normal.

The prolonged P-R interval and ESR returned to normal within a week without specific treatment (Fig 1c). She was discharged, but a week later she was readmitted with pain and swelling in her right ankle, followed, over the next two weeks, by joint symptoms in her right knee and then right shoulder. The development of migratory polyarthritis with previously elevated ESR and prolonged P-R interval on ECG aroused the suspicion of possible RF. This diagnosis was supported by the finding of a raised ASO titre of 400 units. The joint symptoms were successfully treated by Ibuprofen. Salicylates were not given to this patient due to marked improvement in her joint symptoms while on Ibuprofen. She was also advised to carry on penicillin prophylaxis for at least 5 years. On review two months later this lady was in good health with normal ESR, ECG and without any joint symptoms.

Discussion

This lady eventually fulfilled one major and two of the minor Duckett Jones criteria for the diagnosis of RF; that is, fleeting polyarthritis, prolonged P-R interval on ECG and raised ESR. This diagnosis is also supported by the evidence of recent streptococcal infection, as

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Fig 1.

demonstrated by raised ASO titre. The diagnosis in this case was delayed by several weeks due to the lack of sufficient diagnostic features in the early stages of her illness. It was not until the fleeting nature of her joint symptoms came to light quite late in the course of her illness, that the suspicion of RF was aroused.

Symptoms of vague ill health with raised ESR and heart block on ECG may be a presenting feature of several conditions, e.g. SLE, sarcoidosis, scleroderma, bacterial and viral illnesses. In the presence of joint symptoms it is also important to exclude rheumatoid arthritis, Reiter's syndrome, ankylosing spondylitis in adults and Still's disease in children (2).

It is important to make this diagnosis, even in retrospect, for the sake of giving penicillin prophylaxis to these cases. This reduces future streptococcal sore throat and carditis to which these patients are highly susceptible.

Salicylates and corticosteroids have been used successfully in the treatment of RF to alter the course of fever and arthritis, but their role in preventing carditis and its late sequelae remains uncertain (2, 3). Theoretically, non-salicylate NSAIDs should be as effective as salicylates in the treatment of RF, but to date there are no published trials to prove this case. Treatment by salicylates was not considered in this case as her joint symptoms improved on Ibuprofen alone.

Heart failure in RF should be treated by diuretics and digitalis. Bed rest is recommended in the presence of severe arthritis and carditis, as judged by the presence of Carey Coomb's murmur, pericardial rub, effusion on

echocardiogram, ST and T wave changes on ECG and cardiomegaly on chest X-ray. Although this patient had a prolonged P-R interval, she had no other features of carditis on ECG or on clinical grounds.

Residual streptococcal infection should be eradicated with adequate penicillin cover and penicillin prophylaxis should be advised to younger patients up to the age of 25 and in adults for up to 5 to 10 years from the time of diagnosis. But in patients where carditis is the presenting feature, a life-long prophylaxis is favoured by some (4). This patient has been advised 5 years of penicillin prophylaxis, taking into account her overall clinical picture. The response to treatment is judged by improvement in general condition of the patient, along with return of ESR/CRP values to normal levels.

Improvement in living conditions and early use of antibiotics in URTI cases were thought to be reasons behind the significant drop in the incidence of RF. However, a number of recent reports of outbreaks of RF cases from the United States in middle class families with easy access to medical facilities, raise some interesting questions regarding the epidemiology of this disease (5, 6, 7). Appearance of new and virulent mucoid strains of streptococci and genetic susceptibility of individuals are some of the explanations put forward for these outbreaks (3).

This single case report does not signify the resurgence of RF as seen in the United States in recent years. It does, however, highlight the need for the primary care physicians to be aware of the possibility of incomplete forms of this disease which go on to develop the

complete picture later. Polyarthritis may be a presenting feature or may follow later in the course of this illness, as seen in this case.

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