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Acute Rheumatic Fever with Erythema Marginatum-Case Report

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Abstract
Acute rheumatic fever (ARF) is a multisystem disease resulting from an autoimmune reaction to infection with group A streptococcus. Carditis (clinical/subclinical), Arthritis (polyarthritis, mono arthritis, arthralgia), Chorea, Erythema marginatum, Subcutaneous nodules are the classical clinical manifestations. Incidence of ARF is decreasing even in developing countries and the manifestations like erythema marginatum is extremely rare(less than 5%).

INTRODUCTION

Acute rheumatic fever (ARF) is a sequela of streptococcal infection—typically following two to three weeks after group A streptococcal pharyngitis—that occurs most commonly in children and has rheumatologic, cardiac, and neurologic manifestations. The incidence of ARF has declined in most developed countries. Diagnosis rests on a combination of clinical manifestations that can develop in relation to group A streptococcal pharyngitis. These include chorea, carditis, subcutaneous nodules, erythema marginatum, and migratory polyarthritis.We presented this case because erythema marginatum is extremely rare manifestation of acute rheumatic fever.

CASE REPORT

13 yr old school going boy with no significant past medical or surgical history with low socio economic status from costal margins of kerala presented with fever for one week and joint pain for five days. He gives history of sore throat and difficulty for swallowing 2 wks back and the symptoms resolved without any treatment. Sore throat was not associated with any features of viral infection. He was asymptomatic for two weeks and developed high grade continous fever. The joint pain started in left ankle and two days later the pain migrated to left knee and right elbow and wrist. Pain was associated with swelling of left knee and limitation of range of movement of all affected joints. The skin lesions appeared over the front of chest and abdomen. The lesions were serpigenous in fashion, length ~5cm, breadth ~3cm with raised edges, central area was pale and was not itchy.

Within 12 hrs of hospital stay the skin lesions changed their position and size from left side of trunk to right side. The skin lesions disappeared with in 24 hrs after starting treatment with aspirin. The pain of joints and fever subsided within 48 hours after starting treatment with aspirin.

On examination his pulse rate was 110/min, regular. BP-110/80 mm Hg in right arm in supine position and temperature was 101F. There was

mild pallor, lymph node enlargement in left inguinal and cervical area, they were mobile, tender, non matted, firm consistency

There was tinea infection over back of neck and pectus carinatum in general examination.

There was tenderness over left knee and fluctuation with restriction of movement.

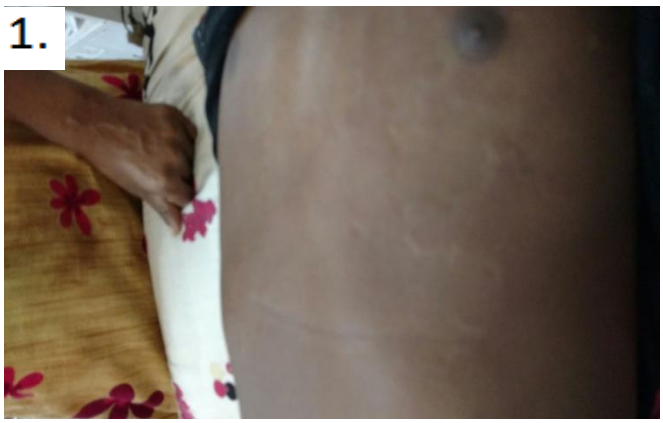
The skin lesions were plaque, erythematous to skin coloured serpigenous passion, varying size over time seen mainly in trunk and abdomen sparing peripheries and face.

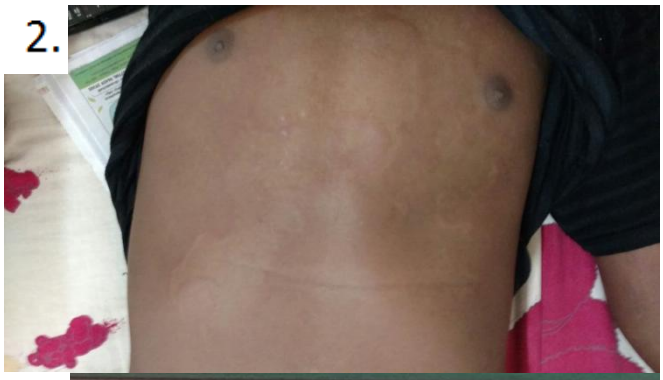
Jugular venous pressure was elevated with prominent "V" wave. cardiovascular system showed soft systolic murmer at apex of grade 2/6 without any radiation, LV S3 was present. There was no cardiomegaly.

Investigations showed haemoglobin 10.6, with hypochromic microcytic picture in peripheral smear and iron deficiency anemia in iron study. ESR was 90.CRP was positive. liver function tests, renal function tests, serum electrolytes, random blood sugar,chest x-ray was normal.ECG showed sinus tachycardia, PR interval was normal.ASO was positive with 390 iu. Echo showed mild MR,TR with features suggestive of rheumatic carditis. however due to the presence of lymphadenopathy excision biopsy was done from cervical lymphnode to rule out other causes. lymph node biopsy was suggestive of reactive inflammatory changes only.

We arrived at diagnosis of acute rheumatic fever with carditis and according to jones criteria

Patient was started on aspirin, pencillin-v and diuretics. Pain and fever subsided with in 48 hrs and the features of heart failure disappeared after one week of treatment. Patient was sent home after three weeks of hospital stay and kept under close follow up for assessing the progression of cardiac involvement with Echo cardiogram





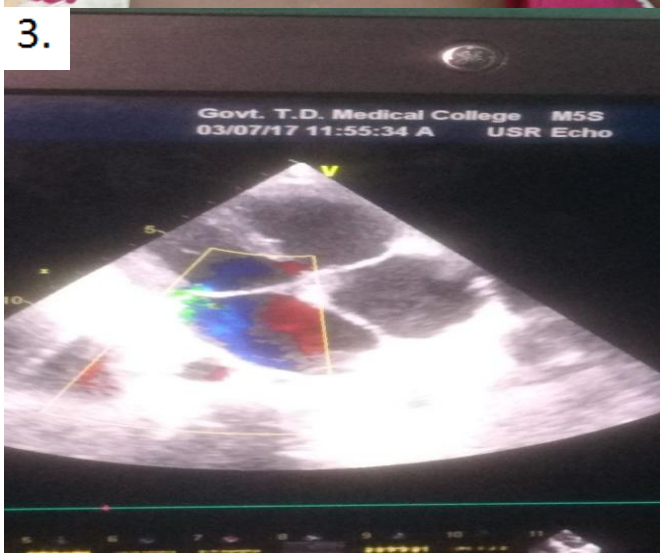


Fig 1&2-Erythema marginatum
Fig 3-echo cardiogram showing mild MR,TR

Discussion

ARF and RHD are diseases of poverty. They were common in all countries until the early twentieth century, when their incidence began to decline in industrialized nations. This decline was largely attributable to improved living conditions, which resulted in reduced transmission of group A streptococci. The introduction of antibiotics and improved systems of medical care had a supplemental effect.

ARF is mainly a disease of children of age 5–14 years. Initial episodes become less common in older adolescents and young adults and in persons age >30 years. By contrast, recurrent episodes of ARF remain relatively common in adolescents and young adults. There is no clear gender association for ARF, but RHD more commonly affects females

Some human leukocyte antigen HLA-class II alleles, particularly HLA-DR7 and HLA-DR4, appear to be associated with susceptibility, whereas other class II alleles have been associated with protection (HLA-DR5, HLA-DR6, HLA-DR51,HLA-DR52, and HLA-DQ)

The most widely accepted theory of rheumatic fever pathogenesis is based on the concept of molecular mimicry, whereby an immune response targeted at streptococcal antigens (mainly thought to be on the M protein and the *N*-acetylglucosamine of group A streptococcal carbohydrate) also recognizes human tissues.

There is a latent period of ~3 weeks (1–5 weeks) between the precipitating group A streptococcal infection and the appearance of the clinical features of ARF. The exceptions are chorea and indolent carditis, which may follow prolonged latent periods lasting up to 6 months

Usually, a latent period of approximately 18 days occurs between the onset of streptococcal pharyngitis and the onset of acute rheumatic fever (ARF). This latent period is rarely shorter than 1 week or longer than 5 weeks. The first manifestation of ARF is a very painful migratory polyarthritis. Large joints such as knees, ankles, elbows, or shoulders are typically affected. Often, associated fever and constitutional toxicity develop. The most common form of joint involvement in ARF is arthritis, objective evidence of inflammation, with hot, swollen, red, and/or tender joints, and involvement of more than one joint (poly arthritis).

Poly arthritis is typically migratory, moving from one joint to another over a period of hours. ARF almost always affects the large joints—most commonly the knees, ankles, hips, and elbows—and is asymmetric. The pain is severe and usually disabling until anti-inflammatory medication is commenced.

Sydenham chorea (rapid, irregular, aimless involuntary movements of the arms and legs, trunk, and facial muscles) was once a common late-onset clinical manifestation but is now rare. Acute attacks usually resolve within 12 weeks.

The classic rash of ARF is *erythema marginatum*, which begins as pink macules that clear centrally, leaving a serpiginous, spreading edge. The rash is evanescent, appearing and disappearing before the examiner's eyes. It occurs usually on the trunk, sometimes on the limbs, but almost never on the face.

Subcutaneous nodules occur as painless, small (0.5–2 cm), mobile lumps beneath the skin overlying bony prominences, particularly of the hands, feet, elbows, occiput, and occasionally the vertebrae. They are a delayed manifestation, appearing 2–3 weeks after the onset of disease, last for just a few days up to 3 weeks, and are commonly associated with carditis.

Heart involvevement in ARF is usually pancarditis and mitral valve is almost always involved.

Diagnosis of ARF is based on modified jones criteria

Diagnosis of acute rheumatic fever, modified Jones criteria, 2015[*]

A.	For all patient populationswith evidence of preceding group A streptococcal infection (other than chorea)	
	Diagnosis: initial ARF	2 major or 1 major plus 2 minor manifestations
	Diagnosis: recurrent ARF	2 major or 1 major and 2 minor or 3 minor
B.	Major criteria	
	Low-risk populations[a]	Moderate- and high-risk populations
	Carditis[b] (Clinical and/or subclinical)	Carditis[b] (Clinical and/or subclinical)
	Arthritis (Polyarthritis only)	Arthritis (Monoarthritis or polyarthritis or polyarthralgia[c])
	Chorea	Chorea
	Erythema marginatum	Erythema marginatum
	Subcutaneous nodules	Subcutaneous nodules
C.	Minor criteria	
	Low-risk populations[a]	Moderate- and high-risk populations
	Polyarthralgia	Monoarthralgia
	Fever (≥38.5°C)	Fever (≥38°C)
	ESR ≥60 mm/h and/or CRP ≥3mg/dL[d]	ESR ≥30 mm/h and/or CRP ≥3 mg/dL[d]
	Prolonged PR on ECG (for age) (unless carditis is a major criterion)	Prolonged PR on ECG (for age) (unless carditis is a major criterion)

ARF: acute rheumatic fever; CRP: C-reactive protein; ESR: Erythrocyte sedimentation rate

[*]Prepared by utilizing the reference number 10.
[a]Low-risk populations are those with ARF incidence ≤2/ per 100 000 school- aged childrenor all-age rheumatic heart disease prevalence of ≤1/per 1 000 population per year.
[b]Subclinical carditis is pathological echocardiographic valvulitis.
[c]Polyarthralgia should only be considered as a major manifestation in moderate- to high-risk populations after exclusion of other causes. As in past versions of the criteria erythema marginatum and subcutaneous nodules are 'stand-alone' major criteria. Additionally, joint manifestations can only been considered in either the major or minor categories but not both in the same patient.
[d]CRP value must be greater than upper limit of normal for the laboratory. Also because ESR may evolve during the course of ARF, peak ESR values should be used.

Treatment of ARF include symptomatic treatment with aspirin. Aspirin is the drug of choice, delivered at a dose of 50–60 mg/kg per day, up to a maximum of 80–100 mg/kg per day (4–8 g/d in adults) in four to five divided doses till the symptoms are substantially resolved, usually within the first 2 weeks, patients on higher doses can have the dose reduced to 50–60 mg/kg per day for a further 2–4 weeks.

Antibiotic prophylaxis should be used for prevention of further attack on already damaged tissues. If no carditis/RHD, prophylaxis may be continued up to 18 yrs or at least five yrs whichever is longer. In case of documented carditis prophylaxis to be continued at least for 25 yrs.In case of chronic carditis often lifelong prophylaxis required.

Use of glucocorticoids is controversial Many clinicians treat cases of severe carditis (causing heart failure) with glucocorticoids. However, the potential benefits of this treatment should be balanced against the possible adverse effects. If used,prednisolone is recommended at a dose of 1–2 mg/kg per day (maximum, 80 mg), usually for a few days or up to a maximum of 3 weeks

Conclusion

Case reports of ARF are decreasing even in developing countries. Better understanding the natural history of this disease and its rare manifestations like Erythema marginatum is important for early diagnosis, treatment and secondary prophylaxis of this disease.