

Case Report

Subcutaneous nodules: a rare and only presentation of acute rheumatic fever

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ABSTRACT

Acute rheumatic fever (ARF) is a most common sequelae of group a beta hemolytic streptococcus (GABHS). 11yr old adolescent male presented with subcutaneous nodules, which is being rarest manifestation, diagnosed to have ARF- mitral regurgitation with active carditis based on revised Jones criteria. Treated with Injection benzathine penicillin, aspirin and steroids according to IAP protocol. Patient become asymptomatic with resolution of subcutaneous nodules. Here we describe the successful diagnosis and management of a patient with ARF

Keywords: Acute rheumatic fever, Carditis, Subcutaneous nodules, Revised Jones criteria

INTRODUCTION

Acute rheumatic fever (ARF) is most common non-suppurative sequelae of group a beta hemolytic streptococcus (GABHS) infection.¹ There are various serotypes present in the environment of which Serotypes-M type 1,3,5,6,18,29 are responsible for ARF.² These serotypes have more affinity towards cardiac valves, sarcolemma, brain and joint causing ARF. ARF with cardiac valve involvement is the important sequelae which has prevalence of >1000 cases per 100,000 population in endemic region.²

Poor socioeconomic status has association with high incidence of ARF which ranges from 100-200 per 100,000 population worldwide in 5-15 years old age group.^{1,3} Pathogenesis of ARF is mainly because of molecular mimicry and immune mediated action against heart valves, sarcolemma, brain, joint.² Diagnosis of ARF is done by revised Jones criteria-2015. There are 5 major (carditis, polyarthritis, erythema marginatum, subcutaneous nodules and chorea) and 4 minor criteria (arthralgia, fever, elevated acute phase reactants-ESR,

CRP, prolonged PR interval) with evidence of recent GABHS infection. For diagnosis of ARF 2 major or 1 major, 2 minor criteria with GAS infection for initial attack is required. In ARF joints, skin, subcutaneous tissues, brain and heart are most commonly involved.⁴ Patients will need only symptomatic treatment during episode for involvement of skin, joint, subcutaneous tissue and brain. But involvement of heart has significant complications like valvular heart disease hence necessitates secondary prophylaxis.^{5,6}

Subcutaneous nodules being rarest manifestation of ARF it is usually missed during diagnosis. Subcutaneous nodules with incidence of 0-10% considered as a rare manifestation of acute rheumatic fever and significantly correlates with RHD.^{2,7} Subcutaneous nodules are firm, painless nodules of approximately 0.5-1 cm usually present along extensor surfaces of tendon near bony prominences in upper limb, lower limb, and in the scalp.² In high-risk population like in Chamarajanagar, state of Karnataka where late manifestation of ARF is increasing one should consider the diagnosis of ARF in patients with subcutaneous nodules. We are presenting a case who had subcutaneous nodules as the only symptoms.

CASE REPORT

A 11-year-old preadolescent male child brought with complaints of fever with painless multiple swelling over extensor surface of upper limb, lower limb and scalp of 2 weeks. There was no h/o joint pain, rashes, palpitations, chest pain, dyspnea, abnormal movements of limbs. No significant past history, family history, developmentally appropriate for age, immunized according to national immunization schedule. On examination child was conscious, oriented and comfortable at sitting posture.

Pulse rate was 134 bpm, regular rhythm, normal volume and character, no radio-radial or radio-femoral delay, respiratory rate was 24 cycles/min, BP was 112/60 mmhg in right and left upper limb at supine position and 116/70 mmhg in right and left lower limb, with Spo2 98% at room air. Head to toe examination showed subcutaneous nodules of about 2×2 cm in occipital region and also over dorsum of left foot near 2nd toe of about 1×1 cm, over right dorsum of hand and around elbow ranging from 0.3-0.5 cm as shown in Figure 1 and 2.



Figure 1: Subcutaneous nodules on scalp.



Figure 2: Subcutaneous nodules in the forearm.

On cardiovascular examination, inspection showed hyperdynamic precordium with apex beat at 5th intercostal space 1 cm lateral to midclavicular line i.e., shifted downwards outwards. Palpation showed hyperdynamic apical impulse at 6th intercostal space 1 cm lateral to midclavicular line and systolic thrill at mitral area. Auscultation in mitral area showed soft S1 with grade IV pansystolic murmur better heard in left recumbent position, radiating to axilla best heard with diaphragm of stethoscope, same murmur was heard in tricuspid area with lesser intensity. Grade II ejection systolic murmur heard at aortic area.

Investigation showed elevated ESR of 36 mm/hr with prolonged PR interval of 236 ms. ASLO titers was 350 IU/dl. Since the patient was fulfilling 2 major criteria, he was diagnosed to have rheumatic fever with active carditis with mitral regurgitation in sinus rhythm, not in failure. Chest X-ray showed cardiomegaly with CT ratio 0.6 with normal liver function and renal function. 2D echo revealed severe eccentric MR, LA dilatation with normal biventricular function, thickened AV with mild AR.

Hence patient was started on Injection benzathine penicillin G 6lakh IU deep IM. I/V/O active carditis tab prednisolone 2 mg/kg/day and planned to start aspirin while tapering prednisolone according to IAP protocol. And patient has been counselled regarding benzathine prophylaxis every 21 days. Patient was reviewed after 2 weeks; he was symptomatically improved with reduction in the size of subcutaneous nodules and decreasing intensity of murmur.

Table 1: Revised Jones criteria. The diagnosis of first RF episode required a confirmation of two major criteria or one major and two minor criteria, along with evidence of antecedent group A beta haemolytic streptococcal infection.

Revised jones criteria ⁸	
Major criteria	
Low-risk population	Moderate to high-risk population
Carditis (clinical and/or subclinical), polyarthritis, chorea, erythema marginatum, subcutaneous nodules	Carditis (clinical and/or subclinical), mono- or polyarthritis, polyarthralgia, chorea, erythema marginatum, subcutaneous nodules
Minor criteria	
Low-risk population	Moderate to high-risk population
Polyarthralgia, fever (≥38.5°c), peak esr ≥60 mm in the first hour and/or crp ≥3.0 mg/dl, prolonged pr interval	Monoarthralgia, fever (≥38°c), peak esr ≥30 mm in the first hour and/or crp ≥3.0 mg/dl, prolonged pr interval

RF, rheumatic fever; CRP, C-reactive protein; ESR, erythrocyte sedimentation rate.

DISCUSSION

Rheumatic fever (RF) is the most common acquired heart disease in the pediatric population between the ages of 5 to 15 years affecting quality of life.⁷ Pathogenesis of ARF is autoimmune cross-reactivity because of molecular mimicry following group A beta hemolytic streptococcal infection. GABHS has more affinity towards cardiac valves, sarcolemma, brain and joint.² Diagnosis of ARF is done using revised Jones criteria shown in Table 1.⁸

Our patient met the revised Jones criteria for moderate to high-risk populations (2015), as evidenced by a positive anti-streptolysin O (ASO) titer, carditis as evidenced by echocardiogram results and subcutaneous nodules. Subcutaneous nodules are rare manifestation of ARF; their incidence is reported to be 0%-10%.⁷ Recent studies showed that there was significant association between subcutaneous nodules and carditis.⁹ Poonia et al, reported that subcutaneous nodules usually manifest four to six weeks after initial infection which accompany with severe carditis.¹⁰

Our patient presented with subcutaneous nodules as the only symptoms. On examination patient had multiple subcutaneous nodules with features suggestive of mitral regurgitation with active carditis, the same was confirmed by echocardiography. Hence patient was diagnosed as ARF with carditis and started with benzathine penicillin, steroids and aspirin. Subcutaneous nodules do not require any specific treatment and will resolve when carditis is treated adequately.¹¹ Patient responded well for the treatment mentioned above, improved symptomatically with no complaints. Henceforth we will continue with same antibiotic every three weeks for secondary prophylaxis to prevent valvular complications of heart.

CONCLUSION

There is lack of awareness and inadequate knowledge in ARF, patients presenting with joint pain, palpitations are most common. Whereas in high-risk areas rare presentation like subcutaneous nodules should be searched for since it is also the major criteria for diagnosis of ARF. We report this case for the rare presentation of subcutaneous nodules, we recommend to have high suspicion of acute rheumatic fever in a child presenting with multiple swelling suggestive of subcutaneous nodules so as not to delay the diagnosis of ARF. And we also recommend evaluation to look for carditis for early intervention to prevent the progression of the disease and cardiac complications.

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