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Jaccoud's Arthritis

K.C. Aggarwal
H. Chellani
R. Mehta
S. Gupta

Rheumatic fever is a systemic disease involving joints, heart, pleura, skin and subcutaneous tissue and basal ganglion(l-4). Although, the incidence of rheumatic fever has come down in advanced countries, it still remains **a** common disease in developing countries like India accounting for 30-40% of total heart diseases(l,2). We present here **a** rare manifestation of rheumatic fever Involving small joints of hands leading to a prolonged disability and deformity called as Jaccoud's Arthritis(l). Recognition of this entity is important specially for differentiation from JRA (Juvenile Rheumatoid Arthritis) as long-term streptococcal prophylaxis will be required for the former. Only an isolated case report of this entity is reported from India and that too in adults(2).

From the Department of Pediatrics, Sanjay Gandhi Memorial Hospital, Delhi Administration, Mangolpuri, Delhi 110 093.

Reprint requests: Dr. K.C. Aggarwal, 198, Pundrik Vihar, Opp. 'D' Block, Saraswati Vihar, Delhi 110 034.

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Case Report

An 8-year-old child presented to us with persistent joint swelling in ankle, elbow and small joints of hand mainly metacarpophalangeal and proximal interphalangeal joints of one month duration. History of sore throat, palpitation, drug intake, epistaxis,

rash and dysuria were denied. Past history revealed presence of fever with major joint involvement 2 years back. Swelling lasted for 8-10 days and the child was not investigated during that episode.

On clinical examination, in addition to joint findings, subcutaneous nodules (size 1-1.2 cm) on olecranon processes and extensor surface of hand were noticed. Heart examination revealed tachycardia and grade 2/6 apical systolic murmur. No other systemic abnormality was detected. Presence-of subcutaneous nodule and heart murmur prompted us to investigate the child for rheumatic fever also.

On investigation, throat swab for J3 streptococci hemolyticus was negative. Hemogram showed polymorphonuclear leucocytosis with ESR of 80. Mantoux test was negative. Rheumatoid factor and ANF were also negative. Slit lamp examination of eyes showed no evidence of iritis. ASO titre repeated twice was 381 Todd units. CRP was positive. X-ray chest showed no cardiomegaly. ECG showed a heart rate of 124/minute and PR interval was normal with no rhythm disturbance. ECHO-cardiography: 2D and M Mode showed edema of mitral cusp with no other abnormality. *X*-rays of joints showed no destruction of bony surfaces but soft tissue swelling around the joints were present. Regional osteoporosis of small joints of hands were also seen. On follow up, the patient was treated with penicillin and aspirin in the doses of 100 mg/kg and supportive care. After 12 weeks of hospitalization, the small joints of hands were still swollen and painful inspite of full coverage with aspirin. On follow up $2^{1/2}$ months after hospital discharge, the swelling of the hands had subsided. There were no subcutaneous nodules. Echo-cardiography had revealed slight thickening of mitral valve cusps without any evidence of regurgitation.

Discussion

Presence of three major criteria, namely, arthritis, carditis and subcutaneous nodules, accompanied by laboratory criteria of high ASO and ESR, positive CRP and polymorphonuclear leucocytosis confirmed the diagnosis of rheumatic fever. However, persistence of small joints arthritis of hands for more than 12 weeks with deformity presented an enigma initially.

Joints are involved in 70% of initial attacks of rheumatic fever(l). The arthritis is classifically migratory (flitting) lasting a day or so in one joint and then disappearing and involving the other joints. It classically involves major joints especially knee joint, elbow joint and ankle joint. Temporomandibular and stemoclavicular joint involvement is not described in rheumatic fever. Very rarely it has been described in rheumatic fever with minor or some deformities of hand and feet that can occur which may be persistent and are known as Jaccoud's Arthritis(4,8). The exact pathogenesis of this condition is not clear. Rarely, this condition has also been described in systemic lupus erythematosis(5), urticarial hypocomplementic vasculitis(6), malignancies and in ulcerative colitis(7). However, this entity assumes special importance when associated with valvular involvement and other features of rheumatic fever. Joint deformity is reversible and overall prognosis is good. At times it may be associated with tendinous laxity and elongation of pattelar tendon which was missing in our case. Another uncommon manifestation of rheumatic fever is simultaneous involvement of multiple joints which was also seen in our case.

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Osteogenesis Imperfecta Type II in One of a pair of Twins

Girish Gupta
Praveen Kumar
Anil Narang

Osteogenesis imperfecta type II also called osteogenesis imperfecta congenita or Vrolik's disease is a rare connective tissue disease affecting 1 in 62,000 births(l). A variety of biochemical defects in type I procollagen resulting in disruption of triple helical conformation and procollagen suicide are responsible for the clinical features(2). Some cases are autosomal recessivtf but many are new dominant mutations. A large majority of patients die in early neonatal period or infancy but the incidence in general population is kept constant as a result of new mutations. It has been estimated that the mutation rate in osteogenesis imperfecta is 4×10^{-5} per gene per generation(3). The occurrence of osteogenesis imperfecta in one of a pair of twins would be a rare chance association. On extensive review of literature we could come across only two such reports(4,5). The purpose of this communication is to describe our experience of such a case, emphasize the inclusion of long bone evaluation in the 'routine' antenatal ultrasound examination and raise the management dilemma posed by such a situation.

From the Division of Neonatology, Department of Pediatrics, Postgraduate Institute of Medical Education and Research, Chandigarh 160 012.

Reprint requests: Dr. Anil Narang, Additional Professor, Division of Neonatology, Department of Pediatrics, PGIMER, Chandigarh 160012.

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Case Report

Baby S, second of the twins, female