There is a need for early selective screening of patients with non-specific clinical symptoms and laboratory findings and initiate emergency non-specific treatment as outlined above if mortality and long term neurodevelopment morbidity are to be improved.

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Valvular Heart Disease: Rheumatic or Rheumatoid ?

Amita Trehan
Surjit Singh
N. Ambalavanan
Lata Kumar

Juvenile rheumatoid arthritis (JRA) was first described by Still in 1897. Extra-articular involvement in JRA is well known but cardiac involvement is said to be uncommon. The association of articular rheumatism and rheumatism of the fibrous tissue

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

Reprint requests: Dr. Surjit Singh, Associate Professor, Department of Pediatrics, Post Graduate Institute of Medical Education and Research Chandigarh 160 012.

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of the heart was described as early as 1836 by Jean Bouillaud. The concept of rheumatoid heart disease was finally made by Bywaters in 1960 and gained acceptance(1). We report here a case of chronic arthritis with cardiac valvular involvement.

Case Report

An 11-year-old boy reported to hospital in August 1992 with a history of pain and swelling of joints for the last 3 years. Arthritis had started in the larger joints and subsequently involved the small joints of his hands and feet. There was history of moderate grade fever during exacerbations of his illness. There was no history suggestive of eye involvement, cardiac involvement or skin rash at any time. He had nodules on his scalp and elbows for 3 months prior to coming to hospital. The child had been treated with aspirin and steroids on and off for 3 years.

Physical examination revealed an obese child weighing 42 kg. His heart rate was 88/min, RR 24/min and BP 120/60 mm Hg in the supine position. Subcutaneous nodules were present on both elbows and over the occiput, 1.0-1.5 cm in diameter. Examination of the cardiovascular system revealed normal first and second heart sounds and a pansystolic murmur in the apex radiating to the axilla. A diastolic murmur was heard in the left parasternal area. The respiratory, abdominal and central nervous system examinations were unremarkable. Mild swelling and inflammation was noted around both knees. Ulnar deviation of both wrists along with spindling of the fingers was obvious. The spine and sacroiliac joints were normal.

Investigations revealed a Hb of 92g/L; TLC of 8 x 10^9/L(N 63%, L 35% and E 2%); ESR of 80 mm in 1 hour; positive CRP; ASO of 305 international units (N <244 international units); negative rheumatoid factor; negative antinuclear factor; negative anti DNA antibodies and negative HLA B 27. X-rays of bones and joints showed marked osteopenia of hand and foot bones, and decreased joint spaces in the metacarpo-phalangeal joints of both hands. Fusion of capitate and hamate.on the right side was also seen. Chest X-ray revealed no cardiomegaly. EKG was normal for age. Biopsy of sub-cutaneous nodule was consistent with a rheumatoid nodule consisting of fibrocollagenous tissue, with central areas of fibrinoid necrosis bordered by histiocytes and few lymphocytes. No Anitschkows cells were seen. Echocardiogram revealed evidence of Grade II mitral regurgitation (MR) and Grade III aortic regurgitation (AR) and posterior movement of the anterior mitral leaflet was seen. A diagnosis of JRA (polyarticular type) with cardiac involvement in the form of MR and AR was made. He was treated with aspirin (80 mg/kg/day) and put on regular physiotherapy. On follow up 2 years later he can walk by himself, has had no exacerbations of his arthritis, no cardiac symptomatology and has now been off aspirin for the last 6 months. His subcutaneous nodules regressed after 12 weeks of therapy. The cardiac legions have remained unchanged. However, as rheumatic fever and rheumatic heart disease could not be ruled out penicillin prophylaxis in the form of benzathine pencillin every three weeks was continued.

Discussion

Cardiac involvement in JRA occurs in about 4.7% of all patient and may present as pericarditis, myocarditis or valvulitis. Pericarditis is considered the commonest lesion(2). Valvular involvement in JRA has been limited to anecdotal case reports. The order of involvement of cardiac valves has been mitral, aortic, tricuspid and pulmonary(3). The main valvular lesion described

has been an incompetent lesion which occurs because of granulomata on the valve leaflets. The lesion most frequently symptomatic is one of aortic incompetence(4,5).

Heart lesions in JRA unlike adults are mainly seen in the sero-negative patients having systemic form of the disease(2,4,6). It has occasionally been seen in sero-positive patients mimicking the adult form of the disease(4). Cardiac involvement is usually asymptomatic and may be picked up on routine echocardiography(4,6). It may occasionally present in an acute manner necessitating valve replacement(4,5).

Our patient had a polyarticular JRA with evidence of cardiac involvement in the form of aortic and mitral regurgitation and possible mitral stenosis. However, there are two questions which need to be answered: (i) Could this patient be having rheumatic heart disease with Jacoud's arthritis?; and (II) IS it possible that he has both rheumatic heart disease as well as JRA?

In answer to the first question, the point most favoring a rheumatic rather than a rheumatoid etiology to his heart disease is the multivalvular nature of the heart disease. He has involvement of two valves with a regurgitant as well as a probable stenotic lesion. Multivalvular heart involvement is rarely seen in JRA and is almost always regurgitant in type. Jacoud's arthritis has characteristic ulnar deviation of metacarpophalangeal joints and extension of the proximal and distal interpha-langeal joints. Even in the absence of these deformities, the roentgenographic changes of osteopenia and decreased joint space favor a diagnosis of JRA. This child had subcutaneous nodules which on biopsy were shown to be rheumatoid in nature. Biopsy of such nodules may not always be confirmatory(7) but the large (>1 cm) size and presence for a prolonged duration suggest rheumatoid etiology(8).

We cannot ignore the fact that this child may have co-existing rheumatic heart disease as well as JRA. The prevalence of RHD in our population of upto 2.1 per one thousand school children(9) makes it a likely possibility that this child has two co-existing diseases and it would therefore, be prudent to continue long term penicillin prophylaxis for rheumatic fever.

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Congenital Chylothorax

Murali N.
Atanu K. Jana
Niranjan Thomas
Jeyanthi P.

Congenital chylothorax is a rare cause of respiratory distress in the neonatal period and yet the most important cause of unilateral pleural effusion in the newborn(1). We present a case of chylothorax that was diagnosed antenatally and managed conservatively in the postnatal period.

Case Report

A booked 20-year-old primigravida mother, who had no antenatal risk factors, had a routine ultrasound scan of the abdomen at 34 weeks gestational age. The fetus was found to have an isolated right sided pleural effusion with no evidence of hydrops or other anomalies. She had an uneventful normal delivery elsewhere and the baby developed mild respiratory distress at 6 hours of age. The infant was breastfed immediately after birth and brought to our hospital at 16 hours of age.

On examination, the baby was found to have tachypnea with dullness to percussion over the right hemithorax. Chest roentgenogram confirmed the presence of fluid in the right pleural space. Aspiration of this fluid yielded 35 ml of xanthochromic fluid with disappearance of symptoms. Examination of the fluid revealed a lymphocyte predominance (96%) and elevated protein content(3 g/ dl). Special stains to detect fat globules were negative. Hemoglobin level, total and differential WBC counts, total protein and albumin levels and echocardiography of the heart were normal. The cultures of blood. and pleural fluid were sterile.

Within 48 hours of admission, the infant developed respiratory embarrassment with signs of reaccumulation of fluid in the right pleural space for which an intercostal drain was placed. The fluid aspirated this time was again tested for fat globules. Though the infant had been breastfed during this time, fat globules were not detected. However, the triglyceride level of the pleural fluid was markedly elevated (>100 mg/dl), strongly supporting the diagnosis of chylothorax.

The fluid drainage continued for a further 48 hours when breastfeeding was stopped and the infant was given a skimmed milk formula along with medium

From the Department of Neonatology, Christian Medical College, Vellore 632 004.
Reprint requests: Dr. Atanu Kumar Jana, Department of Neonatology, Christian Medical College, Vellore 632 004.
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