Childhood 'Rhupus' Syndrome

Co-existence of juvenile idiopathic arthritis (JIA) with juvenile systemic lupus erythematosus (jSLE) is termed childhood Rhupus syndrome. Rhupus syndrome is diagnosed when deforming polyarthritis of JIA and symptoms of SLE co-exist along with positive serological markers [1].

We recently diagnosed Rhupus syndrome in two girls, one presented with deforming polyarthritis of bilateral wrist and knee joints for 9 months, and other with arthritis of right knee and ankle for 11 months. First child also had alopecia, malar rash and oral ulcer for last 2 months, and had hypertension, pallor, hepato-splenomegaly and myelitis, without seizures or psychosis. The other child developed malar rash, oral ulcer and skin bleeds 11 months after the onset of arthritis. Initial investigation in first child showed anemia, neutrophilic leucocytosis with raised erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP). Chest *X*-ray, mantoux test, and liver and renal functions were normal. *X*-ray of wrist showed juxtra-articular osetopenia. Ultrasonograpghy (USG) of knee revealed chronic synovitis. Later, she had proteinuria, positive direct coomb's test (DCT), rheumatoid factor (RF), anti nuclear antibodies (ANA), anti-dsDNA with low C3 and negative anticardiolipin and anti-UI RNP antibodies. Renal biopsy showed grade II nephritis. Whereas, initial investigation in second child revealed anemia, raised ESR and CRP with positive rheumatoid factor. Later she developed pancytopenia, azotemia, hematuria, proteinuria and positive DCT, ANA, ds-DNA with low C3, and negative antiphospholipid and anti-UI RNP antibodies. Kidney biopsy showed grade III lupus nephritis. First child was receiving methotrexate, hydroxycholoroquine and NSAIDs for 6 months for joint manifestations. Later she required pulse methyl prednisolone, followed by oral prednisolone once she developed lupus myelitis, autoimmune hemolytic anemia and nephritis. Second child was managed with NSAIDs, methotrexate, and intra-articular steroid at the beginning, required intravenous cyclophosphamide, prednisolone and azathioprine, once she developed lupus.

The girls had childhood Rhupus syndrome. Rhupus arthropathy is postulated to be either articular involvement of lupus, lupus with chronic polyarthritis or an overlap of lupus with JIA [2-4]. Children with Rhupus present with JIA and later develop lupus. In children, asymmetric erosive and or nonerosive involvements are described [5]. Previous reports showed female predominance, onset at around 8 years, polyarticular involvement, non-erosive arthritis and about 4 year of delay in diagnosis of lupus [5]. Both our patients had deforming polyarticular and oligoarticular arthritis with mean age of 9.5 years and delay in diagnosis of lupus for 9 months. Though rare, childhood Rhupus syndrome is to be considered as a differential diagnosis in patients presenting with deforming arthritis.

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Caffeine in a Term Neonate with Apnea

A term neonate with recurrent central apnea was evaluated at our neonatal intensive care unit (NICU). This male neonate, weighing 3800 g, was vigorous at birth following a caesarian delivery at 38 weeks of gestation. He required continuous positive airway pressure (CPAP) support initially for transient tachypnea but subsequently could not be weaned off as he had recurrent apneas. There was no significant antenatal history. The neonate had recurrent respiratory pauses of 10-40 seconds duration, associated