PAINLESS JUVENILE RHEUMATOID ARTHRITIS (A Case Report)

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Introduction

Juvenile rheumatoid arthritis (JRA) is a chronic inflammatory disease affecting an unknown number of children throughout the world. Synonymous with the entity 'arthritis' is the word 'pain' in the minds of many parents and physicians. Control and relief of pain are the major goals in the treatment of arthritis. A variety of neurological syndromes associated with rheumatoid diseases and JRA are known [1]. The presentation of this case is to highlight the rarity of isolated cranial nerve palsy in JRA and to stress the importance of painless JRA.

CASE REPORT

A 11-year-old male child was admitted with acute onset of ptosis and outward deviation of left eye. No other positive relevant history was elicited from parents. Apart from this the child had been having recurrent attacks of painless bilateral knee swelling for last three years. During each episode, both knee joints used to get swollen up. Child denied history of associated pain, fever, skin rash, involvement of other joints, or visual problems. Throughout the three year period, child was never incapacitated and had been attending school regularly. He had not had the benefit of formal investigations. He had been receiving nonspecific analgesics at irregular intervals and on SOS basis as prescribed by family physician.

Clinically he was a well nourished child with normal anthropometry. Vital parameters were normal. Central nervous system examination revealed normal higher functions. Third cranial nerve palsy on left side, i.e. ptosis, divergent strabismus, and a non-reactive dilated pupil were present (Fig 1). Other cranial nerves were normal. There was no motor or sensory deficit. Fundoscopy and slit lamp examination was normal. Examination of joints revealed bilateral effusion and synovial membrane thickening of both knee joints. There was no other evidence of inflammation. Joint mobility was mildly restricted.

Investigations revealed normal blood counts, ESR 36 mm fall 1st hour, radiogram of chest and Mantoux test negative for

tuberculosis, knee joint radiogram showed increase in soft tissue with no bony lesion, and CSF analysis was normal. Immunological profile revealed positive antinuclear antibody, rheumatoid factor and LE cell negative, and IgM levels normal. Synovial fluid analysis showed turbidity, WBC 250/ μ L, RBC 10/ μ L, predominant cell lymphocytes, and culture was sterile. Histopathological findings of synovium were: (i) hyperplastic synovial tissue with finger-like projections; (ii) sub-synovial connective tissue showing intense edema, plasma cells, lymphocytes and histiocytes; (iii) an occasional Russell body seen amidst the inflammatory infiltrate (Fig 2 & 3).

Fig. 1 : Swelling of both knee joints and ptosis of left eye in the patient.

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Fig. 2: Microphotograph showing numerous villous projections of synovial lining. Stratification of synoviocytes and many dilated and congested vessels also seen. Dense inflammatory infiltrate all around the vessels (100x).

Based on clinical and investigation profile a diagnosis of painless JRA was made. Associated 3rd cranial nerve palsy was an interesting association for which exact etiology was not conclusive. CT scan brain was not done due to paucity of funds. Initial management was with aspirin. Complete recovery of 3rd cranial nerve was observed over the next one week. However, in view of poor response in respect of arthritis, management was changed to methotrexate after 3 weeks of aspirin therapy. The response had been good with almost complete resolution of synovial membrane inflammation. The child is under regular follow up with regular visual screening.

Discussion

Pain control in JRA is a major goal in the treatment of arthritis and is emphasized in medical and lay publications. Lack of pain in a child with other evidence of arthritis prevents him from getting the benefit of specific investigations.

The child as reported in the present report was denied the benefit of specific investigations because of lack of awareness on the part of primary care physician. In a similar report of 41 children with painless JRA, Sherry et al could identify certain characteristics that distinguish painless JRA from painful JRA [2]. These were: (i) delay in diagnosis twice as long in children with painless JRA; (ii) lower mean ESR in children without pain; (iii) arthrocentesis being more often performed in painless JRA. These findings have several practical implications. Foremost is the relevance to the risk of asymptomatic iritis, which is associated with pauciarticular JRA, especially in young girls with positive antinuclear antibody. If

Fig. 3: Higher magnification showing the congested vessels and predominantly plasma cell infiltration.

children with painless JRA have no diagnosis for a year, they could have increased risk for untreated iritis during this time [3]. It is also being theorized that children with JRA complain less of pain than do adults with rheumatoid arthritis [4].

Patients with juvenile chronic arthritis can rarely have neurologic disease. In a series of 170 patients with JRA, 8 had meningismus, 3 had seizures, and 1 had lower extremity weakness. Isolated cranial nerve palsy has been reported in only systemic JRA [1]. Isolated 3rd cranial nerve palsy in this child with pauciarticular JRA does not find any reference. Possibility of viral etiology cannot be ruled out. However, it is less likely in view of negative antecedent history and absence of associated neurologic deficit. The message to be conveyed to primary care physician is that diagnosis of JRA should be considered in children with chronic joint swelling even without pain. Treatment and follow-up of these children should include the appropriate index of suspicion for complications such as asymptomatic iritis and joint destruction.

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