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Case Report

Enthesitis related arthritis with systemic features

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ABSTRACT

Systemic juvenile idiopathic arthritis (SJIA), classified as a form of juvenile arthritis, constitutes around 10-20% of all cases of JIA, and is the most common form of arthritis in children. It has no sex predilection and affects children as young as one year of age. Enthesitis occurs in a form of JIA known as enthesitis-related arthritis (ERA), which has a male predilection and is associated with the HLA-B27 allele. Systemic features do not characterize it. We describe a rare presentation of systemic inflammation in ERA in a female child.

Keywords: Enthesitis-related arthritis, Juvenile idiopathic arthritis, Enthesitis, HLA-B27

INTRODUCTION

Systemic juvenile idiopathic arthritis (SJIA) is a type of inflammatory arthritis with a close resemblance to autoinflammatory diseases that affects children under the age of 16 years. Its distinctive features include high-spiking fevers, evanescent rashes, leukocytosis, elevated acute phase reactants, often organomegaly, lymphadenopathy, and serositis. Fever is the most predominant symptom at disease onset, seen in 98% of SJIA patients.[1]

ERA is another subtype of JIA occurring more commonly in males, associated in the majority with human leucocyte antigen B27.[2] The presentation is with asymmetric oligoarthritis or polyarthritis, predominantly of lower limb joints, associated with enthesitis or sacroiliitis. Management includes NSAIDs with physiotherapy. Conventional disease-modifying agents like sulfasalazine and methotrexate may be used to reduce NSAID use and in those with poor prognostic factors and a high inflammatory burden.[2] In patients who are non-responsive to these drugs, biologic DMARDs such as antitumor necrosis factor alpha agents have proven useful.[3,4] Macrophages have been demonstrated to infiltrate the synovial lining of affected joints with spondyloarthritis. Macrophages produce inflammatory cytokines like TNFα, and blocking this cytokine using monoclonal antibodies against TNF helps improve arthritis and enthesitis.[5]

CASE REPORT

An 11-year-old female child, second in birth order, born of spontaneous conception in a non-consanguineous marriage, had fever and joint swellings of 2 months duration. She also had bilateral conjunctivitis at the onset of the disease, causing the treating clinician to suspect Kawasaki vasculitis and evaluate for any coronary involvement, which was absent. Inflammatory markers, erythrocyte sedimentation rate and C reactive protein were elevated. Serology for juvenile forms of inflammatory arthritis was negative-IgMRF and anti CCP, and ANA-IF. Blood smear examination showed leukocytosis and thrombocytosis with anemia. Bone marrow analysis ruled out any infiltrative or infective pathology and malignancy. Hence, suspecting systemic inflammatory arthritis, NSAIDs were added along with low-dose steroids due to moderate disease activity. The child continued to have high spiking fevers; hence, a PET CT scan was done to look for any internal occult

malignancy. PET CT scan showed a significant uptake in entheses alongside joints. HLA B27 was positive. TNFi etanercept was initiated, however, the child continued to have fever spikes, with only marginal improvement in joint disease activity.

Subsequently, IL-6 inhibitor tocilizumab was initiated to control the high systemic and joint activity, after which her condition improved clinically. Systemic features subsided; however, arthritis persisted, and her functional class deteriorated. TNFi was reinitiated in view of initial responsiveness, and her arthritis subsided over a 4-week duration, and she achieved remission.

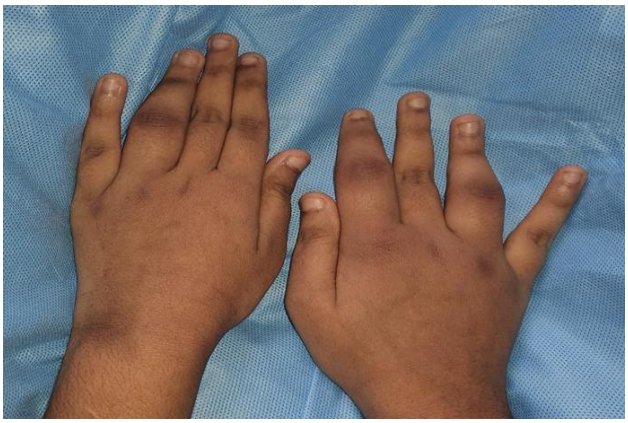

Figure 1: Dactylitis of left 4th, right 2nd and 4th fingers.

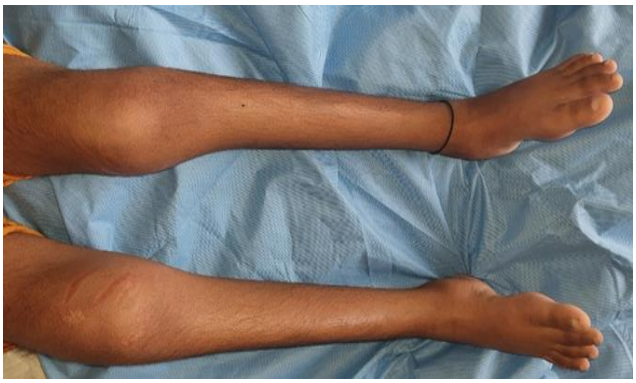

Figure 2: Bilateral knee and ankle synovitis.

DISCUSSION

In a retrospective study by Guo et al it was found that systemic symptoms in the form of fever may be a prominent presenting manifestation in ERA, with about 30% of children with ERA having them. It was also observed that such patients tend to have more active arthritis at presentation.[6] Such an ERA associated with fever also has severe joint disease, which may be refractory to the first-line therapy with NSAIDs and may require the use of glucocorticoids and biologic DMARD therapy.

A high prevalence of HLA-B27 was described in the ERA category of JIA (71%) and a smaller frequency in psoriatic arthritis (50%) the unclassified form of JIA (86.7%).[7] It has also been described in oligoarticular and polyarticular forms, especially in girls.

The clinical presentation in a child with chronic oligoarthritis should entail considering other possibilities, such as infections and post-infectious sequelae, and malignancies. Viral infections like rubella, parvovirus, and dengue can cause acute-onset, generally short-lasting inflammatory arthritis in children, whereas chikungunya virus infection can cause persistent arthritis.[8] Hematological neoplasms can present with joint pains, worse at night, often without much swelling; therefore, it is essential to look at the peripheral blood picture and consider doing a bone marrow examination.[9] In our case, a thorough evaluation had been done to rule out underlying malignancy.

It has been suggested that the presence of ERA is associated with worse long-term outcomes than either the presence of oligoarticular or polyarticular JIA, when followed up for >15 years. Over the longer term, only two-fifths of children attained remission.[10]

CONCLUSION

Though the presence of systemic features raises the alarm to suspect SJIA, a survey for other atypical features for SJIA, like enthesitis, should be done, since on rare occasions systemic features may accompany other types of JIA, such as ERA, which need different treatment approaches.

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