Enthesitis related arthritis: A case report of delayed diagnosis and management

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Abstract
Enthesitis related arthritis belongs to undifferentiated type of Juvenile spondylo arthropathy in JIA subtype of ILAR Classification. Clinical features vary from adult spondylo arthropathy making adult classification criteria inaccurate for paediatric group. Because of significant heterogeneity of this disease phenotype, it has been difficult to propose a classification criteria. JSpA accounts for 20% of JIA with onset usually after 6 yrs with male preponderance (M:F- 7:1). Here we would like to report a case of Enthesitis related arthritis in 11 yr old male which started as Hip joint involvement and later with Lt knee joint and tarsitis. He was managed well conservatively and followed up with DMARDs and supports.

Keywords: Enthesitis, spondyloarthritis, DMARDS, tarsitis, Polyarthrits


Introduction
ERA rarely have sacroiliitis until adolescence and usually present with lower limb arthritis and enthesitis. Articular manifestations of JSpA include hip and peripheral arthritis with an asymmetric oligoarthritis. 1/3 patients will have classical findings of tarsitis. 25% of JSpA have poly arthritis presentation at onset. Enthesitis seen in 60 to 80% of JSpA patients [1]. Early manifestations as enthesitis occur more frequently in children than in adult onset AS. Children may also report vague pain in buttocks, groin and heel. Spinal symptoms are rare, but in sub group where HLAB27+ve boys are likely to progressive to AS in adolescence [2]. JSpA is associated with 6% to 27% of children which typically present with acutely red, painful and photo phobic eye that needs immediate medical attention, if left untreated blindness.

Aortic regurgitation is a main cardiac complication seen in 10% of patients. Subclinical gut inflammation associated with JSpA with findings of as many as 2/3 of patients observed to have changes similar to patients with crohns disease. Arthritis associated with IBD is also seen in children with 2 distinct patterns of joint involvement, one with peripheral arthritis and secondly sacroilitis and spondylitis. Other features like clubbing, periositis, erythema nodosum, pyoderma gangrenosum, osteoporosis and rarely hypertrophic osteoarthropathy are often seen alongside.

Case Report
11 Yr old boy presented to our department with history of pain and swelling of Left knee and ankle pain for last 1 month. Pain in the joints were continuous with no diurnal variation. Patient had history of fever 2 days before the starting of foot pain. There was no history of trauma, gastroenteritis, urinary infection nor eye complaints prior to knee and foot swelling and pain. Patient was ambulatory with an antalgic gait. Active ROM was full on hip, knee and ankle but was terminally painful. There was diffuse tenderness in mid foot. Bowel and bladder habits were normal.

Patient had a history of Lt hip pain 10 months back and was treated in multiple centres before it subsided. Even psychiatric consultation was sought. The reports said patient was treated with...
a diagnosis of Transient bone marrow edema syndrome. He was managed with above knee traction, analgesics and vitamin and calcium supplements. During that time child was having a painful limb. There was no advise of any drugs to be taken for long duration nor any particular follow up advice. After thorough examination we ordered for complete new investigations.


Xray pelvis with both hips AP view (Fig 1) showing normal bony architecture, no evidence of b/l sacroilitis. MRI Hip joint: moderate left hip joint effusion. Focal marrow edema is seen near fovea of head of Left femur head (Fig 2, Fig 3).

With these above positive reports a diagnosis of Juvenile spondyloarthitis was made and patient was started on Indomethacin and methotrexate 7.5 mg weekly with folvate supplements. Patient was given AK skin traction. Active passive ROM exercises, hip abductor extensor strengthening, core strengthening exercises were taught. Patient showed decrease in complaints nearly 2 weeks after start of therapy. Knee and foot pain completely subsided and patient was ambulatory without pain. Patient was referred to ophthalmology and cardiology departments to rule out any extra articular manifestations.

Patient was discharged non symptomatic with Indomethacin, methotrexate 7.5mg, folic acid, Vit D3 and calcium supplements and antioxidant supplements. Patient was asked to review after 1 month for routine follow up and thereafter every 3 months for 1 Yr and every 6m in 2nd Yr. Patient family was counselled about the nature of disease, its course and anticipated complications and progression to adult AS and axial involvement.

Discussion

JSpA is a common under diagnosed JIA. Even in this case the treated physicians neglected the possibility of SpA in 11yr old even though they had a report of secondary bone marrow edema. Patient would have gone for articular damage of peripheral joints if the DMARD was not started timely and patient would have presented with irreversible axial involvement and serious bone deformities after 5 yrs or more. Prevalence of enthesitis-related arthritis is found to be 7.4%, psoriatic arthritis 6.3%, and undifferentiated arthritis 19.7% [6]. Peripheral arthritis especially hip arthritis is the most common presenting features in children [6]. Lower extremity involvement is typically confined to large joints and is asymmetric. Hip arthritis have shown to be an independent risk factor for the development of sacroilitis [8]. 20% of children with JSpA may have “silent” sacroilitis accidently detected on MRI of spine or hip [8].

Extraarticular manifests mainly as unilateral ocular inflammation occurs most frequently in HLA-B27 positive individuals and presents with redness, pain, and photophobia [6]. Gastrointestinal symptoms may also be present in up to 60% of patients with JSpA [6]. Enthesitis is more likely to be a presenting symptom in a child than an adult with the insertions of the patellar and Achilles tendons being the commonest [6].

Plain radiography of the pelvis is the initial imaging modality to look for involvement of the sacroiliac joints. MRI has demonstrated early detection of inflammatory changes, like bone marrow edema of sacroiliac joints [8]. Recent classification schemes propagate the inclusion of MRI-detectable changes into the diagnostic criteria for JSpA [6]. Ultrasound findings of enthesitis include tendon thickening, calcification, or hypo echogenicity and bony erosions at the tendon insertion sites [6].

There is strong evidence of JSpA in genetically predisposed individuals exposed to environmental triggers [6]. Pathogenesis is release of pro-inflammatory cytokines such as TNF-alpha and genetic mutations in the IL-23/IL-17 axis [6]. HLA-B27 accounts for 25% and 2.1% of heritability and genetic mutations respectively.

NSAIDs are the 1st line therapy for both peripheral and axial arthritis. A positive response to NSAIDs pursue further treatment with disease-modifying anti rheumatic drugs (DMARDs). DMARDs like methotrexate and sulfasalazine, have limited applicability in disease control, primarily the axial skeleton. The mainstay of current medical treatment is
anti-TNF therapy and it has shown to limit disease flares, induce disease remission. Children with early axial involvement have a greatest risk for progression to ankylosing spondylitis and are most likely to benefit from early intervention.

**Conclusion**

Main difficulty in adequately treating juvenile SpA is a delay in the accurate diagnosis. Untreated disease can lead to progressive involvement of the axial skeleton and severe deformity and morbidity. Hip joint involvement at younger age greatly increase the risk for hip arthritis and may need THR.

**References**