



## Sacroiliitis in Systemic Juvenile Idiopathic Arthritis – A Rare Clinical Presentation

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### Keywords

Juvenile idiopathic arthritis; Systemic juvenile idiopathic arthritis; Sacroiliitis; Axial disease; Tocilizumab; Infliximab

### Clinical Presentation

Systemic Juvenile Idiopathic Arthritis (sJIA) contributes 10% to 35% of JIA depending on ethnicity and geography [1] and is characterized by high spiking quotidian fever in a child younger than 16 years of age, arthritis along with at least one of these manifestations including evanescent rash, lymphadenopathy, hepatosplenomegaly or serositis, in the absence of other identifiable causes [2]. Wrist, knee and ankle are among the most common joints involved [3]. Sacroiliitis has only and rarely been reported in adult Ankylosing Spondylitis (AS) patients presenting with Adult Onset Still's Disease (AOSD) features [4,5]. Here, we report, for the first time, a case of a sJIA boy developing bilateral sacroiliitis during his course of illness.

A six-year old Chinese boy with no pre-existing medical illness presented with intermittent fever, bilateral ankle arthritis, evanescent rash, hepatomegaly along with progressive weight loss, anemia (Hb 9.3 g/dL), leukocytosis (WBC 15.63/ml × 10<sup>3</sup>/ml) and elevated inflammatory markers (CRP 96.9 mg/L, ESR 83 mm/hr). Intensive investigations for Pyrexia of Unknown Origin (PUO) including septic workup and bone marrow examination were all negative. The ANA showed mixed pattern (1/100 nuclear dots and ≥ 1/800 homogenous) but Extractable Nuclear Antigen test was negative. His rheumatoid factor and HLA-B27 typing were also negative.

The treatment consisted of a short course of oral prednisolone (initial dose of 2 mg/kg/day and subsequently weaned off in 2 months), subcutaneous methotrexate and folic acid. During treatment period his fever and arthritis were resolved. However, his arthritis recurred after two months of stopping oral prednisolone, and then the treatment was switched to intravenous tocilizumab infusion for 2 years. He remained symptom free for 16 months after discontinuing to cilizumab. The flare of disease was triggered by gastroenteritis. It was started with prolonged fever along with an inflammatory type of buttock pain bilaterally, and evolved to polyarthritis involving right elbow, left hip and right first to third metatarsophalangeal joints. The signs of systemic inflammation were also supported by raised inflammatory markers (CRP 56.1 mg/L, ESR 53 mm/hr) and anemia (Hb 10.4 g/dL).

The MRI of sacroiliac joints was consistent with bilateral sacroiliitis with left hip arthritis (Figure 1). He was thus treated again with a short course of oral prednisolone and intravenous tocilizumab infusion leading to the resolution of his joint symptoms with normalization of inflammatory parameters. However, five months later, his bilateral sacroiliitis recurred and infliximab infusion replaced tocilizumab along with a short burst of oral prednisolone. He has since been joint-symptom-free with normalization of all inflammatory markers.

Arthritis is the second most common clinical presentation after fever in sJIA [3]. Even in adults, sacroiliitis was described only in a couple of AOSD case reports in the English literatures [4,5]. Kinsella et al. [5] reported 4 adults with high fever (38.5°C to 39°C) and Inflammatory Back Pain (IBP), but 3/4 had sacroiliitis on X-rays. All improved with phenylbutazone [5]. Another 4 adults presenting with features of AOSD and IBP (3/4 had sacroiliitis confirmed by imaging studies, 2/4 had HLA-B27 positive) reported from Turkey [4]. In 1962, Carter reported a cohort of 202 children

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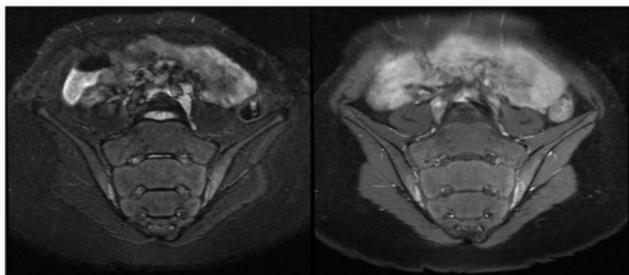
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**Figure 1:** Semicoronal STIR (left) and contrast-enhanced T1w with fat-suppression (right) multiplanar MR imaging of the sacroiliac joints. There is asymmetrical increased T2w signal in the distal aspect of bilateral sacroiliac joints, more on the left than right (left). Synovial and marrow enhancement is also noted (right). There are mild joint effusions. No bony destruction or erosion is noted.

with Juvenile Rheumatoid Arthritis (JRA) with up to 23.7% had probable or definite sacroiliitis diagnosed from X-rays [6]. However, it is unclear as to how many patients with systemic-onset subtype contributed to the cohort. Only one out of 136 systemic-onset JRA children presented with sacroiliitis at the onset of the disease was reported by Behrens et al. [3] from the Pennsylvania Systemic Juvenile Arthritis Registry [3]. To the best of our knowledge, there is no case report on sacroiliitis developed during the course of sJIA.

The immune pathogenesis of sacroiliitis secondary to AS and sJIA may not be identical. Tocilizumab was demonstrated to be effective in controlling both systemic and articular manifestations of sJIA but its efficacy in improving axial disease in AS or juvenile spondyloarthritis was questionable [7,8], and this was demonstrated in our patient that tocilizumab failed to control his axial disease. The effectiveness of anti-TNF including infliximab in axial disease was reported and anti-TNF was included as the first line biologics for axial disease in AS [8], thus anti-TNF may be considered as the first line biologics for sacroiliitis in sJIA.

We report a boy with sJIA who developed bilateral sacroiliitis together with peripheral joint arthritis after his first disease remission. Tocilizumab failed to prevent axial disease recurrence but infliximab thus far controlled the axial disease effectively.

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