

Case Report

Atypical presentation of systemic onset juvenile idiopathic arthritis: a case report

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Received: 05 April 2026

Revised: 04 May 2026

Accepted: 08 May 2026

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ABSTRACT

Systemic Onset Juvenile Idiopathic Arthritis (SOJIA) is an autoinflammatory disorder that can present with a wide range of symptoms, often mimicking infectious etiologies like tuberculosis and other bacterial infections, so atypical presentations can pose a diagnostic challenge. Here, we report a case of a 4-year-old female who presented with high-grade fever for 2 weeks, unresponsive to initial antibiotic therapy. Despite extensive investigations, the focus of infection was not found. Imaging revealed bilateral shoulder joint abscesses, which were sterile, with significantly elevated serum inflammatory markers. Clinical and laboratory findings were consistent with diagnosis of Systemic Onset Juvenile Idiopathic Arthritis (SOJIA) and subsequently corticosteroid and methotrexate therapy were started, to which the child responded.

Keywords: Arthritis, Juvenile idiopathic arthritis, Shoulder abscess, Methotrexate

INTRODUCTION

Systemic Onset Juvenile Idiopathic Arthritis (SOJIA, formerly called Stills disease or systemic juvenile rheumatoid arthritis) is classified as a category of Juvenile Idiopathic Arthritis (JIA) characterized by arthritis with systemic features like persistent fever, rash and prominent visceral involvement including hepatosplenomegaly, lymphadenopathy and serositis.¹

This case underscores the diagnostic challenges and management strategies for SOJIA presenting with atypical features such as joint abscesses, without any symptoms suggestive of joint involvement, which can delay the diagnosis.^{2,3} In this case, the child did not have any features of arthritis like joint pain or restriction of movements. Bilateral shoulder abscesses were accidentally picked up on Computed Tomography (CT) Thorax, which was done to find out the focus of any infection.

CASE REPORT

This 4-year-old female child, born of non-consanguineous marriage, presented with high grade fever (up to 104F) for almost 2 weeks, which was intermittent in nature associated with erythematous rashes in the body, which was transient, occurred mainly during peak of fever. She took oral antibiotic (cefixime) for 5 days, with no improvement. She was diagnosed with MIS-C 6 months ago, treated with steroids. No similar illnesses in family. Her immunizations were up to date and there was no history of weight loss (Weight: 12.5 kg (3rd-15th centile), height: 96.5 cm (3rd-15th centile)). On Examination child was febrile, Other physical examination findings were normal except for hepatomegaly (Liver span 8.5 cm). Initial blood investigations revealed Neutrophilic leukocytosis (Total Leukocyte Count 23000, Neutrophil 64%) with C-Reactive Protein of 314 mg/l and Erythrocyte

Sedimentation Rate (ESR). Chest X-ray, abdominal ultrasound and echocardiography were normal.

Initial diagnosis of Infectious cause (Enteric fever) was kept in mind and was started on ceftriaxone and azithromycin. Blood and urine C/S were negative. In view of persistent fever spikes, antibiotic upgraded to meropenem and repeat blood investigation were done, which showed high inflammatory markers (D-Dimer:1553 ng/ml, Lactate Dehydrogenase (LDH): 496 U/l, Ferritin: 798 ng/ml, COVID Antibody (IgG): 354 BAU/ml; fecal calprotectin: 150 ug/g) suggested the possibility of an autoimmune disease. Antinuclear Antibody (ANA), RF, HLA-B27 were negative.

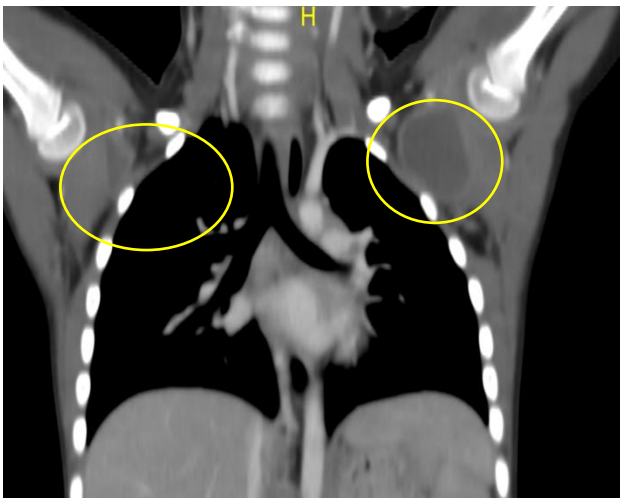


Figure 1: CT thorax showing bilateral shoulder joint abscesses.¹¹

CT Thorax was done as a part of evaluation of unresolving fever, and showed collection in bilateral shoulder joints. Magnetic Resonance Imaging (MRI) revealed Marked subacromial-subdeltoid and sub coracoid bursitis, tenosynovitis of long head of biceps tendon, mild gleno-humeral joint effusion, fluid in axillary recess, left axillary and cervical lymphadenopathy.

USG-guided aspiration of pus was done; which was negative for MTB/AFB stain/Gram stain/fungal cultures/AFB culture. USG guided biopsy from left cervical region Lymph node of size 1.5 cm was done, suggestive of reactive lymphoid hyperplasia. GeneXpert for MTB was reported as negative.

The persistent fever, lack of evidence of an infectious process and joint involvement led to consider the possibility of SOJIA.

She was started on NSAID (Naproxen), fever intensity decreased. Subsequent steroid treatment significantly reduced symptoms and the fever resolved. The child went home with a diagnosis of systemic JIA. She is currently

being treated with Methotrexate and a weaning dose of prednisolone.

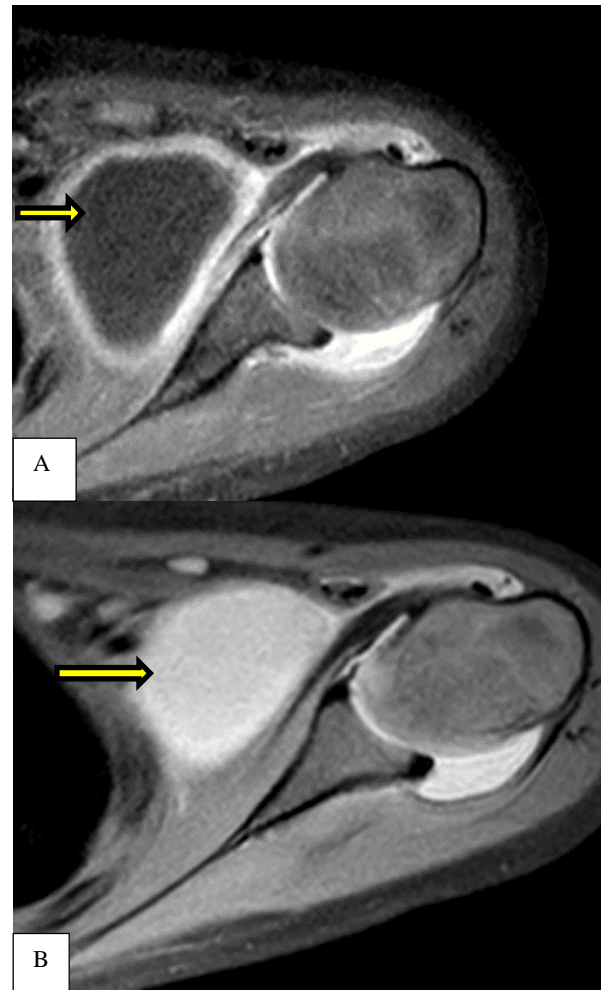


Figure 2 (A and B): MRI scan showing marked subacromial-subdeltoid and sub coracoid bursitis and tenosynovitis of long head of biceps tendon.¹²

DISCUSSION

We report this case to highlight the interesting presentation of SOJIA in a child in the form of fever and bilateral shoulder joint abscess without any other symptoms.³ SOJIA is a subtype of juvenile idiopathic arthritis characterized by systemic symptoms.

According to the International League of Associations for Rheumatology (ILAR), systemic juvenile idiopathic arthritis (sJIA) is defined as arthritis in one or more joints, or arthritis preceded by fever of more than two weeks' duration that is documented to be quotidian for at least three days and is accompanied by one or more of the following features: an evanescent erythematous rash, generalized lymph node enlargement, hepatomegaly and/or splenomegaly, or serositis. Diagnosis is often challenging due to its overlap with infectious diseases. This case highlights the importance of considering sJIA in the differential diagnosis of prolonged fever with

elevated inflammatory markers and joint involvement in children.

CONCLUSION

Early recognition and appropriate management of SOJIA are crucial for improving patient outcomes. This case demonstrates the importance of a multidisciplinary approach in diagnosing and managing complex pediatric cases. SOJIA should be kept in mind if the child presents with persistent fever spikes even without arthralgia.

Funding: No funding sources

Conflict of interest: None declared

Ethical approval: Not required

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Cite this article as: Aadinadh MJ, Bountra A. Atypical presentation of systemic onset juvenile idiopathic arthritis: a case report. *Int J Contemp Pediatr* 2026;13:1029-31.