



## Juvenile Idiopathic Arthritis Complicated with Atlantoaxial Subluxation: A Case Report

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### ABSTRACT

Juvenile idiopathic arthritis is a rheumatological disease that starts before 16 years and involves joints. Many complications such as bone erosions and joint destruction can be seen in the course of the disease. Herein, we presented a case of juvenile idiopathic arthritis complicated by atlantoaxial subluxation followed for 14 years.

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### Introduction

Juvenile idiopathic arthritis (JIA) is a disease that onsets before the age of 16, and manifestations persist for at least six weeks, cannot be explained by any other reason, and can progress with uveitis and arthritis involving one or more joints. It is known that rheumatoid factor (RF) and antinuclear antibody (ANA) positivity can occur, and genetic predisposition (HLA-B27 or HLA-DR4) can be detected. JIA is further categorized as Systemic JIA, oligoarticular JIA (which can be persistent or extended), RF negative polyarticular, RF positive polyarticular, enthesitis-related, psoriatic arthritis,

undifferentiated (none of which can be classified or characterized by more than one group) JIA.<sup>1</sup> Bone erosions and joint destruction, osteopenia and osteoporosis, temporomandibular joint (TMJ) anomalies, mobility problems due to contractures, and ocular complications such as anterior uveitis can be seen in the course of JIA. We aimed to present our case diagnosed in pediatric rheumatology and followed up in adult rheumatology, which was complicated by atlantoaxial subluxation.



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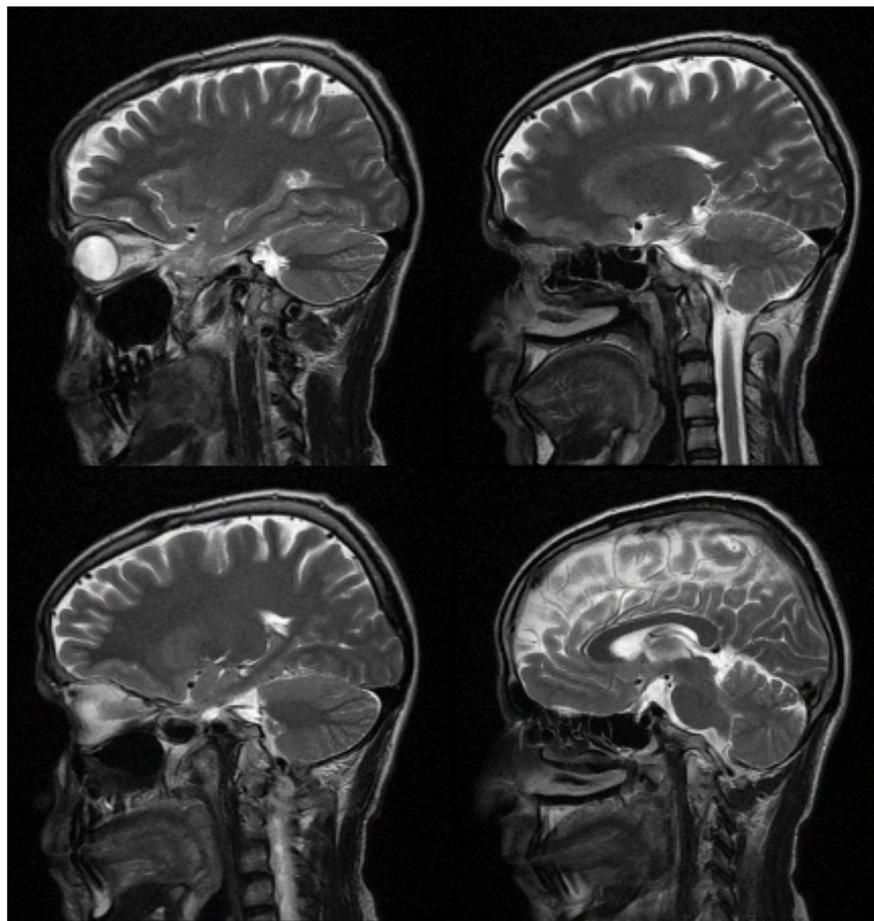
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**Picture 1.** The cranial MR sections showed a “4.9 mm displacement of C2 vertebra odontoid process towards the cranial, in sections passing through the skull base level (basilar invagination). Both cavernous segments are mildly dolichoectatic.”

## Case Report

A 25-year-old male patient, who had been followed up with the diagnosis of JIA for 14 years, described a complaint of dizziness during the control of the adult rheumatology outpatient clinic. The foreign national patient, who was diagnosed with JIA and given steroid, methotrexate and nonsteroidal anti-inflammatory drug (NSAID) treatment after being examined in his home country with a complaint of swelling in the knee and foot joints when he was 11 years old, was transferred to us by pediatric rheumatology at the age of 19. In the examinations performed in pediatric rheumatology, HLA B27 (+) and FMF gene test M694V and E148Q were found as (+). The patient, whose treatment was planned as an

anti-TNF agent, had a history of irregular drug use due to social reasons. The patient had significant mobility restrictions. Findings of arthrosis were observed in both knees and hip joints. After the transfer to us, the patient, who was evaluated in the rheumatology council, was referred to orthopaedics because of arthrosis in both hip joints. Anti-TNF was planned to be started, and he was followed up. In the systemic and neurological examination of the patient who had been using etanercept for five years, no features were found except that he had 2/5 paraparesis (joint movements were completely limited due to bilateral frozen hip). Cranial magnetic resonance imaging (MRI) was performed on the patient. The MRI showed a “4.9 mm displacement of C2 vertebra odontoid process towards the cranial, in sections passing through

the skull base level (basilar invagination). Both cavernous segments were mildly dolichoectatic.” (Picture 1). The patient was consulted to the neurosurgery department with the preliminary diagnosis of atlantoaxial subluxation. Surgical intervention was not considered for the patient who had no neurological symptoms, and control was recommended six months later. The patient was followed up in our outpatient clinic with anti-TNF therapy, and periodic neurosurgery control was recommended for atlantoaxial subluxation.

## Discussion

Cervical joint destruction in patients with rheumatoid arthritis (RA) and JIA may lead to vertebral malalignment (e.g., subluxation), causing pain, neurologic deficit, and deformity.<sup>2-4</sup> In one series, 159 consecutive patients with juvenile chronic arthritis had cervical spine radiographs taken at age 18.<sup>5</sup> In 62 per cent, some changes were noted, including 41 per cent with apophyseal ankylosis and 17 per cent with anterior atlantoaxial subluxation and 25 per cent with atlantoaxial impaction. In another study in one centre, all consecutive patients with JIA were followed into a transition program, and cervical spine radiographs were performed.<sup>6</sup> Of the 57 JIA patients, 65 per cent showed cervical spine lesions, and half had no symptoms.

In conclusion, it should not be forgotten that imaging patients can detect atlantoaxial subluxation findings without symptoms. The young age of the patients is essential in terms of being careful before the surgical procedures that may take place. Although routine imaging is not required in asymptomatic patients, cervical spine imaging is recommended before elective surgery that will require neck manipulation.

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## Conflict of interest

The authors declared that there are no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

## Authors' Contribution

Study Conception: YC, BNC; Study Design: YC, BNC; Supervision: YC, BNC, ED, YP, NL, ZYB; Materials: YC, BNC, AE; Data Collection and/or Processing: YC, BNC; Statistical Analysis and/or Data Interpretation: YC, BNC, ED, YP; Literature Review: YC; Manuscript Preparation: YC, BNC, ; Critical Review: YC, BNC.

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