Early Atlantoaxial Subluxation in Enthesitis-related Arthritis

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Atlantoaxial subluxation is well recognized as a late complication of juvenile arthritis[1]. Early subluxation is uncommon, but it has been reported in 2 patients with juvenile ankylosing spondylitis (AS)[2,3] and in 2 patients with juvenile idiopathic arthritis (JIA)[4,5].

A 12-year-old girl was diagnosed with JIA, enthesitis-related arthritis (ERA) subtype, in March 2012 when she presented with a 1-year history of joint pain and stiffness. She was positive for HLA-B27, and a great uncle had AS. On examination she had arthritis in several proximal interphalangeal joints, the left ankle, right knee, and right midfoot, and inflammation of the left Achilles enthesis. Her cervical and lumbar spine examinations were normal. She was treated with naproxen, prednisone, and methotrexate (MTX). Because of ongoing active disease, her MTX was increased, and sulfasalazine was added in September 2012.

In early December 2012 she reported a 1-month history of mild neck discomfort. Her neck examination was normal. Late that month she presented to her local emergency department with severe acute neck pain and restriction of motion. Her neck was immobilized and she was airlifted to our hospital. Radiographs (Figure 1) and magnetic resonance imaging (MRI; Figure 2) demonstrated severe C1–C2 subluxation. There was no history of major trauma and she was neurologically intact. She underwent posterior C1, C2 fusion with no complications (Figure 3), and etanercept was recommended. At followup after surgery, her parents reported that she had had chiropractic manipulation to her neck to treat neck pain before the subluxation was identified.

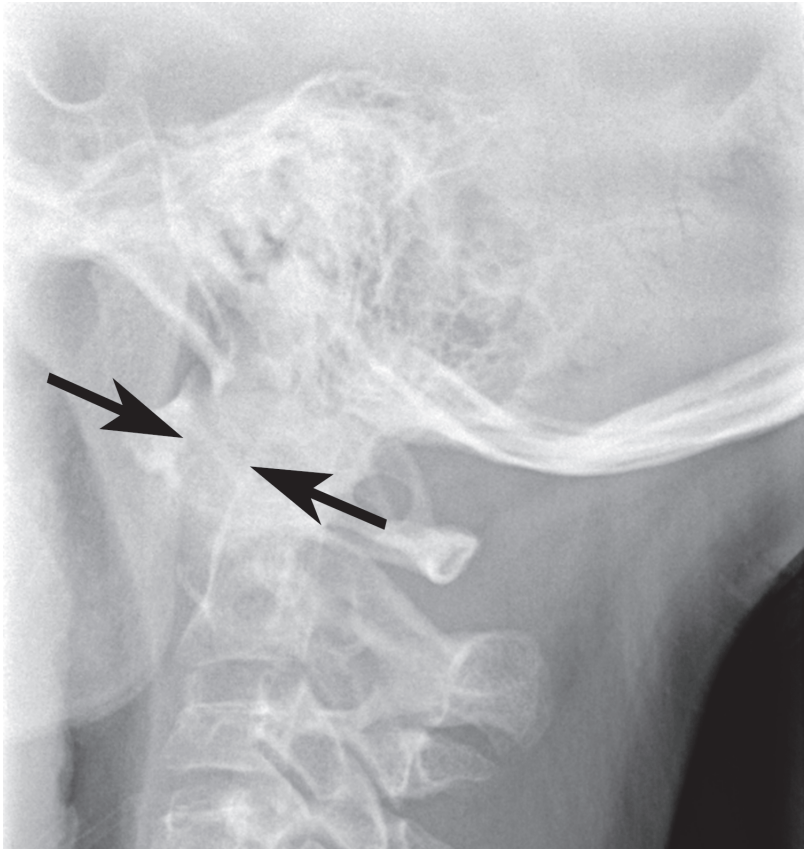

Figure 1. Radiograph demonstrated severe C1–C2 subluxation with increased distance between the anterior arch of C1 and the anterior cortex of the odontoid (arrows).

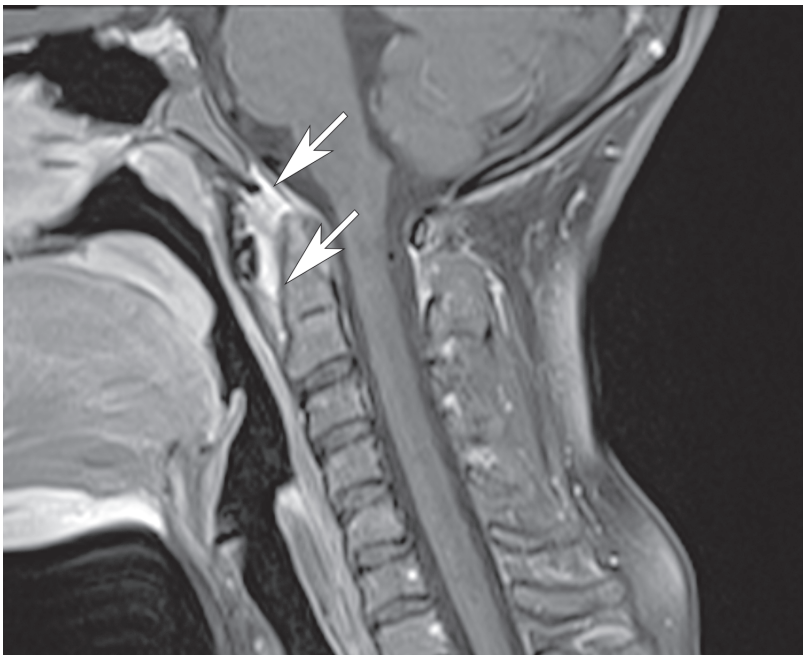

Figure 2. A contrast-enhanced cervical spine magnetic resonance image demonstrated increased separation of the anterior arch of C1 from the dens, measuring 5 mm, with enhancing soft tissue between the tectorial membrane, anterior arch of C1, and around the dens, in keeping with enhancing synovium (arrows).

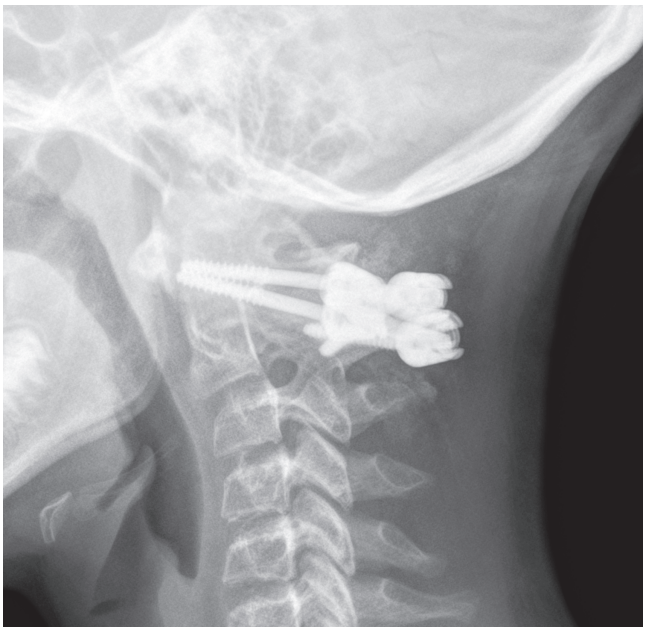

Figure 3. Postsurgery radiograph shows posterior C1, C2 instrumentation and fusion with C1 lateral mass screws and C2 laminar screws.

Clinicians should be aware that atlantoaxial subluxation can occur in children with ERA early in the disease. In our case, the MRI demonstrated synovitis at the atlantoaxial joint, and it is possible that neck manipulation precipitated an acute subluxation. Parents of children with JIA should be cautioned against use of manipulation for neck complaints in their child.

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