

Atlanto-axial subluxation in a patient with polyarticular juvenile idiopathic arthritis: clinical and radiological response to infliximab

Sirs,

Cervical spine is often involved in patients with juvenile idiopathic arthritis (JIA), leading to pain and functional limitations. Atlantoaxial impaction and anterior atlanto-axial subluxation were typical of the upper cervical spine. We describe a case of polyarticular JIA with atlantoaxial subluxation who reached clinical and radiological remission after anti-TNF treatment (infliximab). A ten-year-old boy was referred with a history of chronic urticaria and swelling of both wrists, ankles and first metatarsophalangeal (MTP) of the left foot lasting for 3 months, besides he did complain persistent neck pain. On his first examination, he was growing well. He had bilateral effusions of the wrists, ankles and of the first left MTP. The other joints and systems were normal except for cervical spine restriction. Initial laboratory results revealed normal complete blood count (CBC), urine analysis and complement levels. Immunoglobulin levels (IgG: 1460 mg/dl; IgA: 451 mg/dl) and inflammatory indices (ESR: 82 mm/h; CRP: 3.5 mg/dl) were elevated. ANA was positive (1/640, speckled pattern), anti-dsDNA was negative and rheumatoid factor level was 8 IU/ml (0-14 IU/ml). There were no signs of enthesitis, uveitis or psoriasis. The histocompatibility antigen HLA-B27 was not detected. An inflammatory bowel disease was ruled out by performing an abdominal ultrasound and colonoscopy resulted unremarkable. He was diagnosed as polyarticular JIA. The child underwent MRI of the cervical spine for the presence of neck pain and cervical restriction and an increased atlanto-odontoid distance on cervical x-rays and CT (Fig. 1). MRI showed an atlanto-odontoid distance of 10 mm (normal range 4-5mm) suggesting an atlanto-axial subluxation with an increased transverse atlantal ligament density and an impingement of the odontoid on the upper cervical cord (Fig. 2A). He was treated with steroids (prednisone 0.5 mg/kg) and indomethacin (3 mg/kg) with complete erasing of arthritis except for cervical spine involvement. Considering the potential life threatening complication of atlanto-axial subluxation, infliximab (5 mg/kg) was initiated in addition to steroids at 0.5 mg/kg/day. The child completed the standard infliximab regimen with the initial dose (5 mg/kg infusion at week zero followed by doses at 2 and 6 weeks) followed by the maintenance dose (5 mg/kg every 8 weeks) for nine total infusions. Although the persistent elevation of inflammatory markers (ESR 56 mm/h, CRP 1.6 mg/dl) were consistent with active

Fig. 1. CT at the initial presentation showing an increased atlanto-odontoid distance.

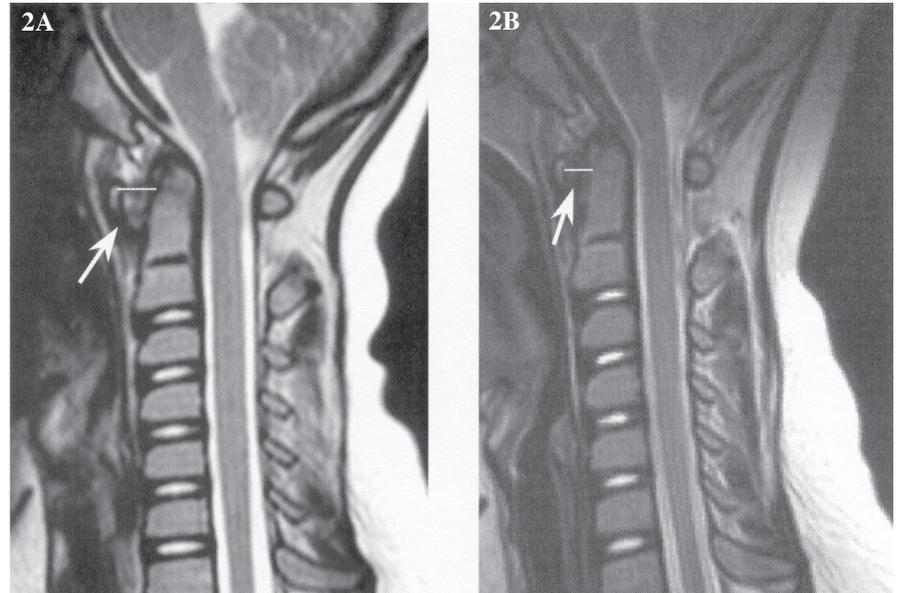
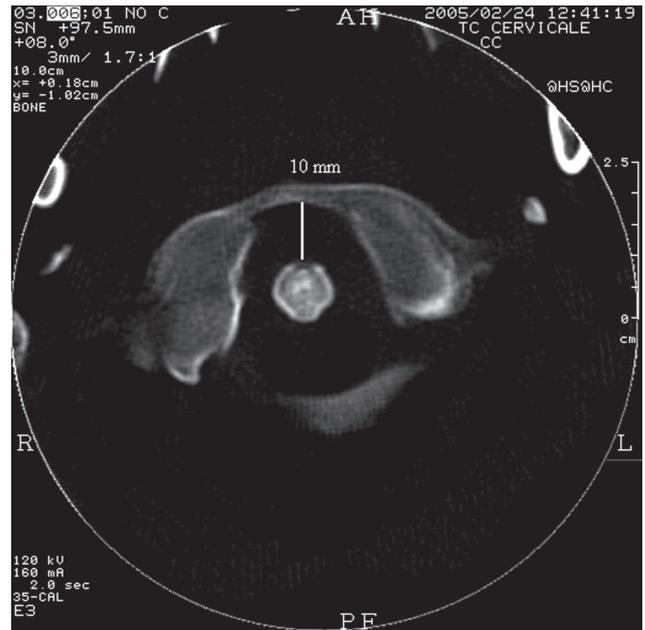


Fig. 2A: Cervical MRI with evidence of atlanto-axial subluxation; 2B: Reduction of atlanto-odontoid distance after 9 infusions of infliximab.

disease, since the third infusion the neck pain and decreased range of motion completely resolved, and steroid therapy was withdrawn. Nevertheless, complete clinical control of the disease was achieved only after the last infliximab infusion, with a normalization of inflammatory markers (ESR 19 mm/h, CRP 0.46) and a follow up MRI of the neck showed a reduction of atlanto-odontoid to a normal value (5 mm) and the disappearance of the transverse atlantal ligament inflammation (Fig. 2B).

Atlanto-axial subluxation is a characteristic radiological abnormality of the axial skeleton in JIA often leading to pain and neck functional limitations (1). Though neurological complications are less likely to develop

in children than in adults (2), an MRI investigation for cervical spine involvement in symptomatic patients is indicated because of the potential life-threatening complications of the lesion.

Although the cervical spine involvement is quite common in JIA; it usually responds to early and aggressive treatment with NSAIDs and steroids but no consensus exists on those subset of patients not responders to conventional therapy.

Recent data show a quick and long lasting response to TNF- α antagonists in adult patients affected with psoriatic arthritis and atlanto-axial subluxation (3). To our knowledge, this is the first case of polyarticular JIA with atlanto-axial subluxation not responding to

standard therapy that showed a complete remission after TNF blocker therapy according to the efficacy of anti TNF medications in the treatment of JIA (4).

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