Atlanto-axial joint involvement as exclusive manifestation of juvenile idiopathic arthritis

Sirs,

Non-traumatic anteroposterior atlanto-axial subluxation (AAS) has already been described in different rheumatic settings such as rheumatoid arthritis (1) and ankylosing spondylitis (2).

Juvenile idiopathic arthritis (JIA) is the most common inflammatory rheumatic disease in childhood and rarely involves the cervical spine at onset especially in children with polyarticular course (3). In these cases, upper cervical spine involvement is usually accompanied by other kinds of joint involvement (4).

We report the case of a child presenting with isolated inflammatory AAS. An 8-year-old girl presented with unresolved painful torticollis for the preceeding 6 months. There was no history of preceding trauma or illnesses, and had no other signs or symptoms. Joints examination was completely normal. Laboratory investigation was normal with no evidence of inflammation; HLA B27 was absent; tests for rheumatic factor (RF) and anti-nuclear antibodies (ANA) were negative.

During this period she underwent a computerised tomography (CT) scan showing an atlanto-axial rotatory malalignment and calcifications in odontoid process; later a magnetic resonance imaging (MRI) scan demonstrated, in a sagittal T2-weighted sequence, an impression of the anterior front of the dura at C1-C2 level with the presence of solid hypointense signal consistent with inflammatory tissue (Fig. 1). An axial TSE T2 fat-suppression sequence revealed subluxation and mild rotation of C1-C2 vertebrae (figure not shown). Questionable fluid and ligaments thickening around the dens were noted. No evidence of fractures, bone destruction, disc protrusion, cervical cord involvement or tumour was found, including images of the posterior fossa. The effusion and calcifications suggested an inflammatory nature of the process and a working diagnosis of oligoarticular JIA with the sole atlanto-axial involvement was contemplated. A trial with monthly courses of methylprednisolone pulses (30 mg/kg/day for 3 days) for 2 months resulted in a temporary resolution of the pain and a partial improvement in head movement. As the limitation of cervical motion persisted, methotrexate (15 mg/mt square weekly) was prescribed. However no significant improvement was achieved at the six-month follow-up.

Because of the peculiar clinical presentation, the persistence of normality of laboratory tests and the very limited response to therapy, the diagnosis of oligoarticular JIA was called into question.

On further follow-up, a routine ophthalmology examination detected a subclinical iridocyclitis in the left eye; considering that presence of uveitis in ANA negative oligoarticular JIA patient is quite uncommon, we performed some laboratory tests aiming to rule out infections (Lyme borrelliosis, Bartonella Hensalae, Herpetic infections, Cytomegalovirus), which supported the autoimmune origin of eye inflammation and further confirming the JIA diagnosis.

Treatment with infliximab was started (6 mg/kg/dose): a concrete improvement on neck stiffness and range of motion, as well as the complete resolution of uveitis, was rapidly obtained by the second infusion.

To our knowledge, this is the first case of oligoarticular JIA involving only the upper cervical spine in absence of other clinical sign of joint involvement.

Acquired chronic torticollis is often the clinical sign of an atlanto-axial rotatory malalignment; its differential diagnosis is extensive including inflammation or infections of the tissues of the upper airway, neck and/or pharynx, traumatic injuries, cervical cord or posterior fossa brain tumour or severe visual disturbance. In order to rule out most of these conditions, MRI and/or CT scan of the brain and cervical spine must be carried out.

In our patient, torticollis was the clinical sign of JIA. AAS has already been described in both rheumatoid arthritis (RA) and JIA. Its incidence varies between 17% to 88% of patients with RA (5, 6), but only in 3% of patients with early disease (7), while among children it seems to be more rare, and mostly associated to polyarticular course JIA and juvenile spondylarthropathies (3). AAS and rotatory fixation has already been described before as the first manifestation of HLA-B27 positive seronegative spondyloarthropathy that went undiagnosed for 11 weeks (8). Similarly, others described acute torticollis as the initial manifestation of systemic onset JIA preceeding other disease manifestations for 2 weeks (9).

Interestingly, upper cervical spine involvement seems to be absent in patients with oligo-articular JIA, probably because it is a less severe and uncomplicated disease with insignificant use of glucocortocosteroids (3).

Indeed, the weak response to treatment, with persistent normality of routinary autoimmune screening, associated with unusualness of joint involvement could lead to review diagnosis; but it was the discovery of uveitis that convinced us that JIA was the proper diagnosis. As a consequence we prescribed infliximab resulting in a favourable response of both uveitis and cervical arthritis (10).

In conclusion, we have described a child with atlanto-axial involvement as the sole articular manifestation of oligo-articular JIA. Our case proves again the importance of routine ophthalmology evaluation to detect acute subclinical uveitis in oligo-articular JIA, and to confirm JIA diagnosis, in atypical cases. In addition, the excellent response of cervical spine involvement to infliximab therapy is also of note.

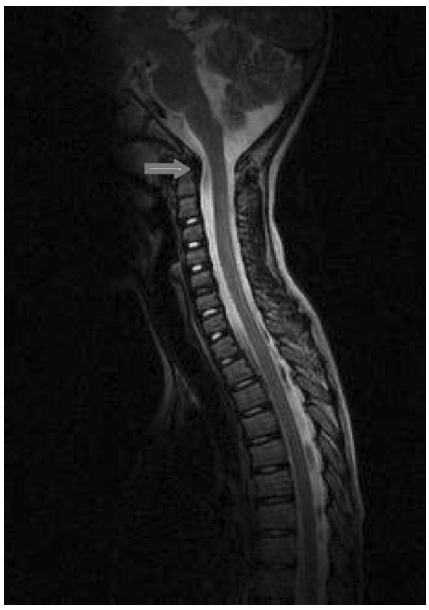

Fig. 1. Sagittal T2-weighted sequence showed at C1-C2 level the presence of solid hypointense signal and the impression of anterior front of dura (arrow).

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