Atlantoaxial rotatory dislocation (AARD) in pediatric age: MRI study on conservative treatment with Philadelphia collar—experience of nine consecutive cases

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Abstract

Purpose Atlantoaxial rotatory fixation and atlantoaxial rotatory subluxation are the most frequent manifestations of atlantoaxial rotatory dislocation (AARD) in pediatric population and are often treated conservatively. The objective of this study is to correlate the changes highlighted on MRI T2-weighted and STIR sequences with the duration of conservative treatment.

Methods We analyzed nine consecutive patients treated surgically between 1 Jan 2006 and 1 Jan 2010 at the Policlinico Umberto I of Rome. All patients underwent cervical X-ray, computed tomography and magnetic resonance imaging (MRI) (T1 and T2-weighted, STIR, angio MRI). All patients were treated with bed rest, muscle relaxants and cervical collar, and radiological follow-up with MRI and cervical X-ray was performed.

Results According to Fielding's classification, we observed seven patients with a type I subluxation and two patients with a type II subluxation. In type I, STIR and T2 sequences showed a hyperintensity in the alar and capsular ligaments and in the posterior ligamentous system, with integrity of the transverse ligament (LTA). In type 2, the hyperintensity also involved the LTA. During the follow-up, MRI showed a progressive reduction until the disappearance of the hyperintensity described, which was followed by a break with orthotic immobilization.

Conclusions MRI with STIR sequences appears to be useful in addressing the duration of conservative treatment in AARD.

Keywords AARS · AARF · Cervical subluxation · Conservative treatment · Rotatory dislocation

Introduction

Pediatric spinal injuries represent the 1–10% of all spinal injuries and the 5% in childhood [1]. Car accidents represent the main cause, followed by domestic accident, particularly relevant in younger age while sports injuries are more frequent in older children [1]. In children younger than 8 years of age, the cervical spine is the region most frequently involved, while in younger children the involvement of cranio-cervical junction is more frequent [1, 19]. This happens because of the different biomechanical characteristics of the pediatric spine, such as: (1) increased mobility and (2) greater susceptibility to distraction and subluxation injuries. Therefore, the biomechanical mechanisms are represented by distraction, hyperflexion, hyperextension forces, and clinical sequelae which may be highly disabling or even lethal. The atlantoaxial rotatory fixation (AARF) and the atlantoaxial rotatory subluxation (AARS) are the most frequent manifestations of atlantoaxial rotatory dislocation (AARD) in children, and conservative treatment has proved to be suitable in many cases, considering the pathological features of these type of injuries. In literature, there is no agreement on the treatment modalities and the timing of conservative treatment. The aim of the study is to correlate the changes highlighted on MRI T2-weighted and STIR images with the duration of conservative treatment with the Philadelphia collar.

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Materials and methods

This prospective study considered all pediatric patients affected by AARD admitted at the Department of Emergency and Acceptance of the Policlinico Umberto I in Rome between 1 Jan 2006 and 1 Jan 2010. All patients were evaluated neurologically and studied at the entrance with RX (A/P, L/L and transoral), CT and MRI (1.5 T, T1-weighted, T2-weighted and STIR sequences and angio MRI). Based on the neuroradiological findings, patients were classified according to the classification of Fielding and Hawkins [3]. All patients were initially treated with bed-rest and muscle relaxants (Pridinolo mesylate 0.2 mg/kg/day) until the spontaneous reduction of the subluxation occurred. A Shanz collar was employed in an early stage, while a Philadelphia collar was preferred after the improvement of the subluxation. All patients were assessed at follow-up with clinical examination and serial MRI studies (T1-weighted, T2-weighted and STIR sequences, sagittal, axial and coronal cuts, angio MRI) at 1, 3 and 6 months from starting treatment, and cervical dynamics X-rays were performed after 6 months. All patients have removed the Philadelphia collar after the disappearance of inflammatory signs showed on MRI (Fig. 1).

Results

Nine consecutive patients (6 males and 3 females, mean age of 5.7 years, range 4–8 years), affected by AARD were admitted at our DEA between 1 Jan 2006 and 1 Jan 2010. The causes of the trauma were: car accident in four cases, school accident in two cases, and domestic accident in three cases. All patients presented with torticollis, neck pain and the cock-robin typical posture. Focal neurological deficits were not present in any case. In all patients, the

neuro-radiological examinations showed an AARS, in particular, seven patients had a type 1 subluxation according to Fielding and Hawkins and two had a type 2 subluxation. In patients with type 1 subluxation, performed MRI showed the integrity of the LTA and hyperintensity of the alar ligaments, articulation involved in the subluxation and the PLS in the T2-weighted and STIR sequences (Fig. 2c, d). In patients with type 2 subluxation, in addition to these findings, there was a hyperintensity of LTA, but without any real interruption. In all cases, within a week of starting treatment, there was a complete disappearance of the cock-robin antalgic posture and the complete reduction of the subluxation seen at the RX control. At 1 and 3 months, the T2 and STIR sequences showed a progressive reduction of the hyperintensity, indicative of a reduction of inflammatory-edematous phenomena of the joint and ligaments involved in the trauma. In five patients, the hyperintensity was not present on STIR and T2 sequences at 3 months and in the remaining four at 6 months (Fig. 3). The normalization of the radiological features was followed by a break with orthotic immobilization. At the dynamic X-rays performed 6 months after the trauma, there was no evidence, in any case, of a delayed instability. At 1 year all patients were symptoms free (Table 1).

Discussion

AARF consists in the complete atlantoaxial dislocation while in the AARS the dislocation is rather incomplete. The AARS is almost always reducible, while the AARF can be reducible or irreducible, and may represent the evolution of an AARS [15]. These are caused in children by rotary component forces that occur mainly because of a trauma. However, they may also occur after head and neck surgery, inflammation of the soft tissues adjacent or of the upper respiratory tract (S. Grisel) and congenital anomalies of the cranio-cervical junction. In particular, in children, there are three important aspects to note: (1) more horizontally oriented facet joints, resulting in a greater range of anterior and posterior movements, (2) wedge-shape of the anterior vertebral bodies, which facilitates their forward slide and (3) low development of the hooked processes that allows rotatory movements. Moreover, we have to add a greater laxity of ligamentous structures and the lack of development of the paravertebral muscles, spinous processes and occiput condyles [4–6, 8, 10, 11, 13, 15, 16, 18, 19]. Everything is important to explain the hypermobility of the pediatric cervical spine and its greater susceptibility to distraction and subluxation injuries. These differences persist at least until the age of 8–10 years, a period in which the osteo-muscular and ligamentous structures take on characteristics

Fig. 1 Diagnostic algorithm

Fig. 2 Plain cervical radiography shows normally aligned C1–C2 complex and a suspected C2–C3 anterolisthesis (a); CT axial scan shows fixed atlantoaxial rotatory subluxation (b); STIR sequence

shows hyperintensity involving C1–C2 articular capsules, especially in the left side (c); MRI T2-weighted imaging shows hyperintensity in the left alar ligament (d)

similar to those of the adult [4–6, 8, 10, 11, 13, 15, 16, 18, 19]. The larger size of the head relative to the rest of the body also contribute to shift the focus of the movements (pivot point) more cranially (C2–C3) than in adults (C5–C6); As a consequence, the cervical trauma below the age of 8–10 years are reflected mainly at the cranial-cervical junction [13]. From a biomechanical point of view, the movements of rotation in the cervical spine depend for 60–80% from the complex C1–C2, with maximal C1–C2 rotation of approximately 70°, and head rotation axis consists of the odontoid [17]. These movements are limited by the alar ligaments and capsular ligaments of the joint C1–C2: alteration of these structures is the anatomical basis of the AARS and AARF. Dvorak et al. [2] studied 43 adult patients with suspected rotary instability as a result of car accidents, and 60% of these showed an increased passive rotary motion at C0–C1 and C1–C2, suggesting insufficiency of the alar ligaments. Up to pathophysiology, the subluxation causes spasm of the paravertebral muscles: the spasm increases the pain and stabilizes the subluxation which causes the spasm. If not treated

promptly, the AARS may evolve into AARF, much more difficult to treat. Fielding and Hawkins [3] have classified the atlantoaxial subluxation in four types: type 1, the most common, consists in a rotation of C1 on C2 without anterior or posterior displacement; type 2, in which there is an anterior displacement of C1 on C2 between 3 and 5 mm; type 3, in which the anterior displacement is greater than 5 mm; type 4, where C1 is displaced posteriorly compared to C2. The possible association to a rotary dislocation to an anterior or posterior displacement of C1 on C2 suggests an alteration of the transverse ligament, in addition to the damage of joint structures and alar ligaments. In our series, according to Fielding and Hawkins's classification, we observed seven patients with a type 1 subluxation and two patients with a type 2 subluxation. From the clinical point of view, all the patients presented with torticollis, neck pain and a characteristic posture in which the head was rotated in one direction and tilted in the opposite direction (cock-robin). All our patients were neurologically intact. Radiologically, the transoral X-ray can show an asymmetry in the separation

Fig. 3 Follow-up 3 months MRI: T2 coronal (a) and STIR coronal (b) sequences and T2 axial (c) and STIR axial (d) sequences show normal signal both in the articular capsules and the alar ligaments 3 months after trauma

Table 1 Distribution of patients' characteristics

Age	Gender	Fielding and Hawkins classification	Hyperintensity (T2, STIR)	Cause of injury	Loss of hyperintensity (STIR, T2), (months)	Delayed instability
5	F	I	Alar ligaments, articular capsules	School accident	3	–
4	F	I	Alar ligaments, articular capsules	Domestic accident	3	–
7	M	II	Transverse and alar ligaments, articular capsules	Car accident	6	–
8	M	I	Alar ligaments, articular capsules	Domestic capsules	3	–
5	M	I	Alar ligaments, articular capsules	School accident	6	–
6	F	II	Transverse and alar ligaments, articular capsules	School accident	6	–
7	M	I	Alar ligaments, articular capsules	Car accident	6	–
5	M	I	Alar ligaments, articular capsules	Domestic accidents	3	–
5	M	I	Alar ligaments, articular capsules	School accident	3	–

between each lateral mass of the atlas and the odontoid peg; this radiological study was performed in all the patients, resulting non-diagnostic in one patient because of the less compliance caused by marked painful symptoms. The cervical latero-lateral X-ray does not allow a diagnosis of AARF and AARS, but it was performed at the beginning of the diagnostic screening in addition to the transoral X-ray to

exclude the presence of other cranio-cervical lesions. This study has not an high accuracy for the AALS and AARF diagnosis, but it can bring out abnormality of the cranio-cervical junction and shape of the arch of the atlas, resulting from anterior displacement of a lateral mass and posterior of the other (Fig. 2a) [13]. In all patients, latero-lateral X-rays were performed to execute the following measurements to

identify a cranio-spinal junction instability: ADI, LADIs and Wackenheim line. If associated with anterior displacement of C1 on C2, the latero-lateral X-ray shows an increase of ADI (n.v. <5 mm in the pediatric population). This finding was highlighted in the two patients with a type 2 subluxation (even if remained <5 mm), in which the rotatory dislocation was associated with an anterior displacement, while in the remaining seven patients none of these three indices was altered. The cervical CT scan, associated with the 3D reconstructions, has a high accuracy in the diagnosis of AARF and AARS. It allows to highlight the rotation of C1 on C2 with the resulting anterior displacement of the lateral mass in one side, posterior displacement of the contralateral mass and the lateral displacement of the odontoid peg due to a trauma of the alar ligaments and locked facets (Fig. 2b). On the other hand, Maas and colleagues [7] have shown that the performance of CT scan in patients with typical cervical posture “cock-robin” can cause frequent errors of interpretation, often resulting in a non-diagnosis of AARF and AARS, and suggested a major use of CT scan with total anesthesia. In our nine patients, CT scan helped us to see the rotatory subluxation of C1 on C2 and to exclude any cervical subaxial injuries. In our experience, however, the most useful examination has proved to be the MRI despite the absence of focal neurological signs, as it has allowed the detailed analysis of the ligamentous structures of the CS junction. The damage to the ligamentous structures is highlighted by a hyperintensity on T2-weighted sequences as a result of an exudate typical of the inflammatory response in the injured tissues (Fig. 2d). The hyperintensity seen on T2-weighted sequences is not easily distinguishable from adipose tissue that often surrounds and is present in the context of the capsule-ligamentous structures; that is why, up to our experience, the use of STIR sequences or T2-weighted sequences with fat signal suppression is necessary (Fig. 2c). STIR sequences were preferred over fat-suppressed T2-weighted sequences, because they give more homogeneous fat signal suppression and a better definition of the inflammatory edema [12]. In this study, the signal changes were highlighted in the joint capsules, PLS, alar ligaments and, if associated with anterior displacement (type 2 according to Fielding and Hawkins) in the transverse ligament; in this case, in particular, lesions can be localized in the context of the ligamentous structures or in their insertion points on the bony structures (Fig. 2c, d). These alterations, present in the acute phase, were no longer evident in the MRI control 3 months after trauma in five patients, and were not evident after 6 months in the remaining four patients (Fig. 3). We hypothesized that the progressive reduction until the disappearance of such signal changes could be a sign of restoration of total integrity of the capsule-ligamentous structures of the CS junction. From the clinical point of view, the decision to remove the Philadelphia collar was taken in all cases on the

basis of this radiological evidence. MRI was also helpful to provide a non-invasive angiography of the vertebral arteries, which may be interested in this type of pathology in their course within the transverse foramina: in one patient we have shown a reduction in size of a vertebral artery. This finding remained asymptomatic and resolved with the restoration of the physiological position of the cervical spine. As described in the literature, this pathology is treated with rigid type Halo-vest orthosis, for periods not exceeding 6 months; there is also no evidence in the literature of a minimum period of duration of the treatment for the resolution of the pathophysiology [9, 14]. However, our nine patients were treated conservatively in a Philadelphia collar (until the disappearance of the hyperintensity in the STIR and T2 sequences), muscle relaxants and bed rest (until the disappearance of torticollis and the recovery of the physiological position of the column cervical). The choice of orthosis was made taking into account: its effectiveness, the type of capsule-ligamentous abnormalities seen on MRI (no real capsular ligamentous solutions of continuous), the young age of the patients and their poor compliance to a conservative treatment such as using Halo-vest. Bed rest was very helpful in the initial step, because it eliminates the axial load on the C1–C2 complex and seems to be the main factor involved in spontaneous reducing of fixation subluxation [15]. Muscle relaxants administration resulted to be useful in all patients to reduce the amount of muscle spasm and pain. In all our patients, conservative treatment has resulted in a reduction of subluxation–fixation within a week after admission. Once the clinical status improved, all patients were discharged with the indication to wear the Philadelphia collar until the complete regression of the altered signal seen during the follow-up with MRI. The radiological follow-up with MRI in all patients showed a progressive decrease and the disappearance of the hypersignal of the alar ligaments, capsular structures and the transverse ligament, as well as the correct positioning of the odontoid peg and lateral masses of the atlas (Fig. 3). The cervical X-ray in flexion–extension, with the measurement of the indices of instability of the cranio-cervical junction (ADI, Ladis and Wackenheim line), has excluded the presence of delayed instability in all patients.

Conclusion

MRI, and in particular T2-weighted and STIR sequences as parameters for the evaluation of the AARD healing process in children, seems to be very useful in addressing the duration of conservative treatment, indicating precisely the improvement of the abnormalities occurred in the capsular ligament. The use of the Philadelphia collar seems to be adequate in relation to the alterations present in the MRI and should be preferred over such an orthosis as Halo-vest,

since it has proved to improve the compliance of young patients. All this is extremely important, especially in pediatric patients, because it enables us to reduce the time necessary for immobilization in a cervical orthosis.

Conflict of interest None.

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