

Early Diagnosis and Management of Combined occipitoatlantoaxial rotatory fixation, Case Report and Review of the Literatures

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Abstract

Background Data: atlantoaxial rotatory fixation (AARF) although rare, is a well-known condition to most spinal surgeons. Occipitoatlantoaxial rotatory fixation (COAARF) is a condition that may develop as a compensatory event following AARF making diagnosis more difficult. Most cases of COAARF present late making their management a difficult task.

Purpose: to demonstrate a neglected case of COAARF

Study Design: case report

Patients and Methods: we report a 9 years old girl with neglected COAARF following minor trauma. 7 months after trauma we could not reduce its rotation operatively.

Results: delayed diagnosis of COAARF precluded its reduction

Conclusion: COAARF can result from relatively minor trauma and should be suspected in children with persisting neck pain and decreased neck movements. Early diagnosis of COAARF is important and facilitates its management. MS and 3D CT-scan is the corner stone in its diagnosis. (2013ESJ053)

Key Words: combined occipitoatlantoaxial rotatory fixation, neck trauma, children

Introduction

Atlantoaxial rotatory fixation (AARF) although rare, is a well-known condition to most spinal surgeons. Occipitoatlantoaxial rotatory fixation (COAARF) is a relatively rare condition.^{4,10} Both conditions are common in pediatric age group and are secondary to minor trauma. COAARF is a condition that may develop as a compensatory event following AARF.^{11,17} Patients with COAARF has less rotated and tilted head than those

with AARF only, because of counter occipitoatlantal rotation.¹⁶ This often makes the accurate diagnosis difficult and late and hence complicates the issue of management. Most cases of COAARF present late making their management a difficult task. Very few cases have been reported in the literatures.^{3,4,5,7,10,11,13,16,19} In this study we reported a case of COAARF after minor trauma and discuss the problem of diagnosis and management of such cases.

Case Report

History:

In this study we present a 9 years old girl presented with some neck pain for 7 months. Her condition started after falling while playing at school. She suffered severe neck pain with typical cock robin posture. At that time she was neurologically intact and had limited painful neck movement. Her plain radiograph was assessed as normal by a non-specialist and she was managed by anti-inflammatory drugs and cervical collar. After that she had physiotherapy for some time. During that time her pain gradually decreased and also her neck posture regained near normal alignment. As her neck pain persisted, she sought further medical advice and further imaging was done. At that time she received more physiotherapy and manipulation but without any response. She was referred to our service almost seven month after here trauma with some neck pain.

Examination:

On examination, she was neurologically intact with minimal neck toricollis. She had some limitation of her neck movements. She had 25 degree of neck flexion, 30 degrees of both side lateral neck rotation, and 35 degrees of both side lateral neck bending. She had no evidence of any systemic illness of upper respiratory problem.

Radiology:

Here plain radiographs showed evidence of minimal cervical rotation without any clue to a specific diagnosis. (Figure 1,A) Also her MRI was inconclusive regarding the diagnosis though it demonstrated no sign of cord compression or any cord signal change. (Figure 1,B) Her CT-scan axial views showed typical 53 degrees clock-wise rotation of the atlas (C1) in relation to the occiput (O) above and axis (C2) below. Both O, C2 appeared in neutral position in relation to other spine. (Figure 1,C). whereas, CT-scan coronal views showed almost the same data viewing C1 lateral mass rotation in relation to O, C2 (Figure 1,D)

Management:

She was submitted for clinically and radiologically monitored progressive skull tong traction up to 10 kilograms for one week without response. After that operative reduction was undertaken through bilateral lateral cervical approach to undo fusion

between lateral mass of the upper cervical spine, which proved ineffective intraoperatively. She was left for monitored traction for another week without response. Then she was managed with Philadelphia collar for further 3 month after that her pain was completely relived and her neck position was near normal, with some acceptable limitation of neck movement.

Discussion

Rotatory subluxation of the atlantoaxial joint was described for the first time by Corner⁶ in 1907. Persistent subluxation causing torticollis was termed as atlantoaxial rotatory fixation (AARF) by Wortzman and Dewar²⁰ in 1968, followed by Fielding and Hawkins⁹ in 1977. AARF is relatively uncommon condition that requires a high suspicion index for diagnosis.^{1,2} Correct diagnosis with X-ray is usually difficult because the patient has severely bended head and stiff neck in many cases. Mandibular and occipital bone can be overlapped and makes it difficult for diagnosis. The same principles apply to COAARF with more difficulty because of the double rotation.

AARF although rare, is a well-known condition to most spinal surgeons. COAARF is a more relatively rare condition and most spinal surgeons are not familiar with.^{4,10} Both conditions are common in pediatric age group and are secondary to minor trauma. COAARF is a condition that may develop as a compensatory event following AARF.^{11,17} Patients with COAARF has less rotated and tilted head than those with AARF only, because of counter occipitoatlantal rotation.¹⁶ So it is often difficult to diagnosis accurately. Most cases of COAARF present late making their management a difficult task.

Only 7 pediatric patients with OAARF have been reported in the literature since it was first published in 1959 up to this year.^{3,4,5,7,10,11,19} since the case published by Fusco et al,¹⁰ in 2011, only two cases have been published by Lee et al and Kim et al^{13,16} this year 2013, both patients were above 18 years old.

COAARF seems to be a compensatory result of uncorrected AARF as patients chronically attempt to correct for their abnormal head position and neck pain.¹¹ It could be caused by the violent motion in rotation and traction that caused a compensatory contralateral subluxing of occiput-atlas onto the

Figure 1. a: plain radiograph lateral view showing inconclusive position of atlas vertebra, b: MRI lateral image showing no significant abnormality, c: CT scan axial cuts showing clock wise rotation of the atlas vertebra in relation to adjacent occipital condyles and lateral mass of axis vertebra, d: CT scan coronal reformats document again same finding as c.

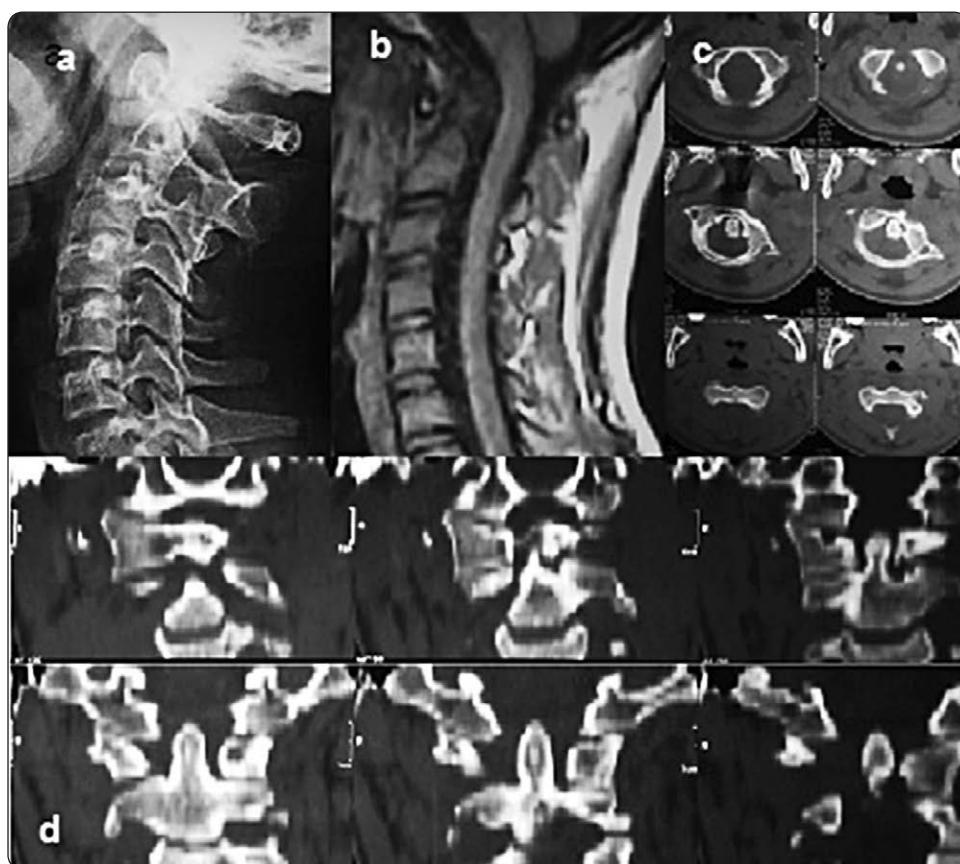


Table 1. Reported Patients of COARF

Authors /Yr	Age /yrs	Duration /mos	Head Position	Cause	Treatment
Washington, 1959	11/F	4	neutral	pharyngitis	traction & Calot jacket
Clark et al.,1986	16/M	8	obvious cock-robin	trauma: fall from horse	traction, O–C2 arthrodesis, halo bracing
Altongy & Fielding, 1990	9/F	2	obvious cock-robin	trauma: break dancing	traction, C1-2 arthrodesis, halo bracing
Cowan & Inglis, 1996	13/M	9	obvious cock-robin	postsurgical traction	C1-2 arthrodesis
Hettiaratchy et al., 1998	13/F	9	neutral	upper respiratory infection	traction, C1–2 transarticular fixation, halo vest
Bouillot et al., 1999	17/F	<0.25	slight head tilt	trauma: chiropractic manipulation	traction, O–C2 arthrodesis
Fusco et al., 2011	8/F	9	neutral	JIA	traction, O–C2 in situ fixation/fusion, collar
Lee et al., 2013	19/F	<0.25	slight head tilt	Trauma: RTA	traction, C1–2 fixation, halo vest
Kim et al., 2013	51/M	192	obvious cock-robin	Trauma: fall Diplegic CP	Traction, 0-C5 fixation/fusion, halo vest
Present Case	9/F	7	neutral	Trauma: fall	Failed closed, open reduction, in situ C1-2 arthrodesis

initial atlas-axis subluxation⁵ It has been also demonstrated that 64.5% of compensation to a vision-forward head position in chronic AARF occurs via the occipitoatlanto joints and only 35.5% occurs via the subaxial spine.¹⁷ Established Occipitoatlantoaxial rotatory subluxation further progress to fixation through certain mechanisms. These mechanisms include ligamentocapsular contractures,⁹ fibrous formations within the synovial joint,¹⁸ inflamed adherent synovial surfaces, osseous union between C-1 and C-2, and abnormal facet deformities.¹²

Data of all reported cases in the literature including this case is summarized in table 1. COAARF has been reported so far in 8 children aged 9–17 years and two adults following either traumatic injuries (fall from a horse, break dancing, and chiropractic manipulation), infections (pharyngitis and upper respiratory infection), or ear surgery. All but two have reported long duration of symptoms (range 2–9 months, except one case reported 16 years), resulting in late or even missed diagnosis. Reported head positions ranged from neutral (N=4), to a slight tilt (N=2), to a cock-robin position (N=4).

Both plain radiography and MRI provide inconclusive data regarding the accurate diagnosis of this condition.^{1,2} Although MS-CT scan overcomes this problem and is considered the imaging of choice for this condition, yet 3D-CT provides accurate diagnostic image and gives a global profile for the rotated spine.^{10,14} Although MS-CT and 3D-CT-scan improve the diagnosis, still normal atlantoaxial rotation makes it a challenge. Functional/dynamic CT has been recommended by many authors for diagnosis of a fixed rotation before the decision of surgery is taken and to exclude normal atlantoaxial rotation^{9,15,17}

External immobilization and closed reduction have been universally ineffective in OAARF.^{3,4,5,7,11,19} Closed reduction for OAARF have been attempted using either; manual reduction under anesthesia,⁴ Halter traction with cervical bracing for 6 months,¹⁹ and Gardner-Wells traction or halo immobilization.^{3,4,5,7,11,13,16} The decreased rates of success in closed reduction with delayed treatment of AARF has been reported by some investigators.^{2,17} This applied also for COAARF, where delayed management was the cause for failure of closed

reduction and even open reduction in some cases¹⁰ including our case. When thoroughly reviewing the previous reports, we noticed that it was not clear that rotation was reduced prior to surgery in all patients. It looks that most patients were fixed in situ after trial of closed and open reduction. (Table1)

For irreducible AARF, most surgeons have tried open reduction via posterior midline approach. Crockard and Rogers⁸ used successfully the extreme lateral cervical approach to break the atlantoaxial lateral mass fusion bilaterally in AARF. We used the same technique in our COAARF case, but it was also ineffective. This can be explained by the chronicity of our case (7 months history), and development of further adhesions across double counter rotation of the occipitoatlantoaxial joints. Bilateral breaking of adhesions of the occipitoatlantoaxial joints resulted in decortication of these joint which initiated auto-fusion and improved neck pain of our patient through the following 2 months with Philadelphia collar. That was our justification for abandoned fixation and continues conservative therapy trying to preserve the present neck motion in our child.

The above data support our conclusion that there should be physician awareness of the COAARF to help early diagnosis in order to pick up cases at early stage where reduction and management is easy feasible target rather than at a late stage where reduction might be a challenge. The simple and effective screening image we recommend for any case of torticollis would be axial CT scan.

There is no established guideline in the current literatures for the management of COAARF as the condition is extremely rare. Reviewing the published cases we noticed that two types of therapy have been proposed. All reports proposed first closed reduction followed by open reduction when closed reduction fails. Following this step they were divided into two groups of management. The first group^{4,5,10,13} proposed O-C2 fixation and arthrodesis. The second group^{3,7,11,16} proposed C1-C2 fixation and arthrodesis and Halo vest. The second group believed that normal flexion-extension function of the occipitoatlantal joint was worth preserving. As the condition is extremely rare and so far only 10 patients including our have been reported in the literature it is hard to extract an evidence based algorithm for COAARF management.

Conclusion

COARF can result from relatively minor trauma and should be suspected in children with persisting neck pain and decreased neck movements. Physician awareness of the condition is mandatory for early diagnosis. Early diagnosis of COARF is important and facilitates its management. MS and 3D CT-scan is the corner stone in its diagnosis.

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الملخص العربي

التشخيص المبكر وإدارة جمع الاستدارة الثابتة للفقرات العنقية الأولى والثانية مع مؤخرة الرأس،
تقرير حالة ومراجعة للأدب

خلفية البيانات: على الرغم من ندرة الاستدارة الثابتة للفقرات العنقية الأولى والثانية فهي حالة معروفة لمعظم
جراحي الفقرات. شوكي. في حين جمع الاستدارة الثابتة للفقرات العنقية الأولى والثانية مع مؤخرة الرأس هو حالة
قد تحدث كتعويض بعد استداره الفقرات الأولى والثانية مما يجعل التشخيص أكثر صعوبة. معظم هذه الحالات
تأتي في وقت متأخر مما يجعل إدارتها مهمة صعبة

الغرض: عرض حالة مهمة لجمع الاستدارة الثابتة للفقرات العنقية الأولى والثانية مع مؤخرة الرأس

تصميم الدراسة: تقرير حالة

المرضى والطرق: هذا تقرير حالة مهمة لفتاة تبلغ من العمر ٩ سنوات تعاني من جمع الاستدارة الثابتة للفقرات
العنقية الأولى والثانية مع مؤخرة الرأس في أعقاب صدمه طفيفة. بعد ٧ أشهر من الصدمة لم يتمكن ارجاع
الاستداره جراحيا

النتائج: تأخر التشخيص يمنع العلاج المناسب

الاستنتاج: يمكن أن تنتج هذه الحالة عن صدمة طفيفة نسبيا، وينبغي ان يشتهب في الأطفال الذين يعانون من
استمرار آلام الرقبة وانخفاض حركات الرقبة. التشخيص المبكر مهم ويسهل إدارته هذه الحالة. الأشعة المقطعية
متعددة المحاور ثلاثية هي حجر الزاوية في التشخيص.