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Case Report

Nontraumatic atlantoaxial rotatory subluxation in adults: Report of two cases

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ABSTRACT

Background: Nontraumatic infectious atlantoaxial rotatory subluxation (AARS) is rare and less frequently encountered in adults versus children. We utilized a stepwise approach to treat two adults with nontraumatic infectious AARS and summarized the relevant literature.

Case Description: Two patients, ages 35 and 66, presented with classic clinical and imaging findings for infectious nontraumatic AARS. Here, we summarized the management for these two patients along with the literature.

Conclusion: Nontraumatic infectious AARS in adults requires prompt X-ray diagnosis and timely application of traction to minimize neurological deficits. MR/CT imaging next offers critical information regarding whether operative stabilization is warranted.

Keywords: Atlantoaxial, Cock-robin, Grisel syndrome, Nontraumatic, Subluxation

INTRODUCTION

Nontraumatic atlantoaxial rotatory subluxation (AARS) associated with an infectious or inflammatory etiology is often managed conservatively with traction or bracing. It is mostly seen in the pediatric age group. The severity of subluxation is usually based on the Fielding and Hawkins classification system [Figure 1].[4,9] Insufficiency of the transverse ligament can result in vertebral deformity with cord compression which leads to neck pain, a locked "cock-robin" position of the head and neck, dysphagia, and/or dyspnea.[7] Here, we present two adults, ages 35 and 66, both with nontraumatic infectious AARS, and highlight how their management differs from that for patients in the pediatric age group.

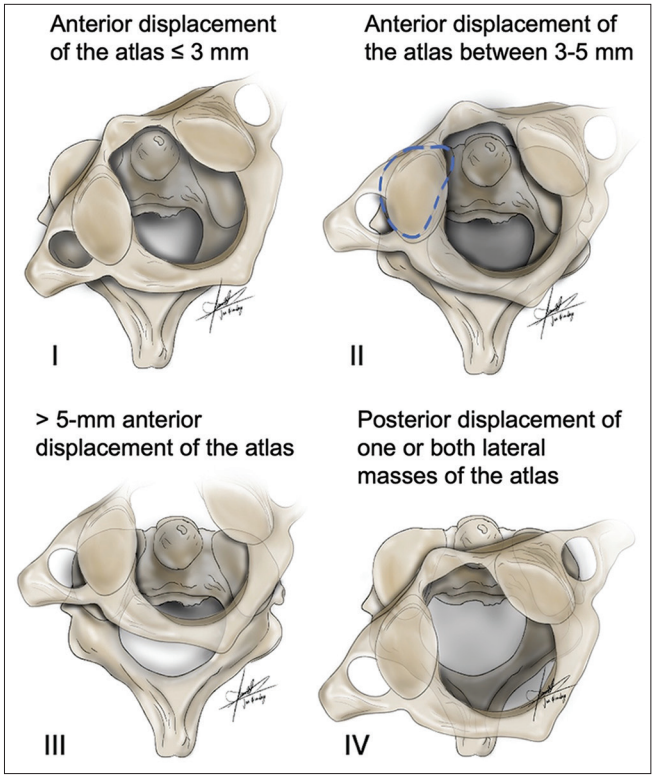

Figure 1: Fielding and Hawkins classification of atlantoaxial rotatory subluxation.

CASE DESCRIPTION

Case 1

A 66-year-old female presented with 2 months of dysphagia, neck pain, dysarthric speech, and severe torticollis with the classic "cock-robin" position for several weeks' duration [Figure 2a]. She was initially diagnosed with a retropharyngeal abscess (i.e., elevated white blood cell (WBC) count of 14,000/mm^3, erythrocyte sedimentation rate of 34 mm/h, and C-reactive protein level of 226.2 mg/L). However, when magnetic resonance imaging (MRI) and X-rays showed severe C1/C2 cord compression with anterolisthesis, infectious AARS was diagnosed [Figures 2b and c]. She was reduced by sequential traction (up to 10 lbs) followed by halo application 5 days later [Figure 2d]. Repeat MRI and contrast-computed tomography (CT) confirmed infectious AARS and led to an occiput-C5 posterior spinal fusion with a C1 laminectomy [Figures 2e-h]. Postoperatively, she remained on empiric antibiotics for 6 weeks at which point all cultures were negative. The 3-month postoperative MRI demonstrated stable reduction of the C1-C2 subluxation with adequate cord decompression [Figure 2i].

Case 2

A 35-year-old male with a history of intravenous drug abuse presented with 1 week of fevers, upper neck pain

Table 1: Select case reports of nontraumatic infectious AARS in adults in the literature.

References	Age, sex	Cause	Time to diagnosis	Fielding and Hawkins classification	Reduction methods	Outcome of reduction	Surgical treatment
Dhaon *et al.* 2003[1]	20, M	Tuberculosis of occiput	5 months	Type I	Halter traction for 1 week, then four-post collar until 4 weeks after pain resolution	No recurrence at 1.5 years	——
Doshi *et al.* 2007[2]	78, M	Skull base osteomyelitis	asymptomatic	Type III*	Abx for 6 weeks	Worsened subluxation	ORIF (not specified)
Ferdinandov *et al.* 2021[3]	48, M	Retropharyngeal abscess, IV drug use	1 month	Type III	——	——	C1-2 fusion
Guleryuz *et al.* 2002[5]	30, F	Pharyngitis	1 week <2 months	Type I	Abx for 6 weeks; Collar for 6 months	No recurrence at 1 year	——
Ishikawa *et al.* 2020[6]	67, F	Pyogenic MSSA spondylitis	2.6 months	——	Abx; traction reduction and halo stabilization for 2 weeks	——	Occiput-C3 fusion
Kerolus *et al.* 2015[7]	71, M	Culture-negative endocarditis	<1 month	Type III	Abx; Aspen collar for 8 weeks	Worsened AARS	C1-4 fusion
	35, F	Molar extraction, otitis media, SA bacteremia	7 weeks	Type III*	Abx; Aspen collar for 12 weeks	Worsened AARS	C1-2 fusion
Panopalis *et al.* 2005[8]	51, M	SA bacteremia	2 days	Type I*	Abx	No recurrence	——
Yamazaki *et al.* 2008[10]	26, M	Retropharyngeal infection	>2 months	Type III*	Abx; halo vest for 6 months, then Philadelphia collar	Recurrence	Occiput-C3 fusion

AARS: Atlantoaxial rotatory subluxation, Abx: Antibiotics, F: Female, IV: Intravenous, M: Male, MSSA: Methicillin-sensitive *Staphylococcus aureus*, ORIF: Open reduction and internal fixation; SA: *Staphylococcus aureus*, *Fielding and Hawkins classification was not explicitly stated by the authors but inferred from imaging parameters

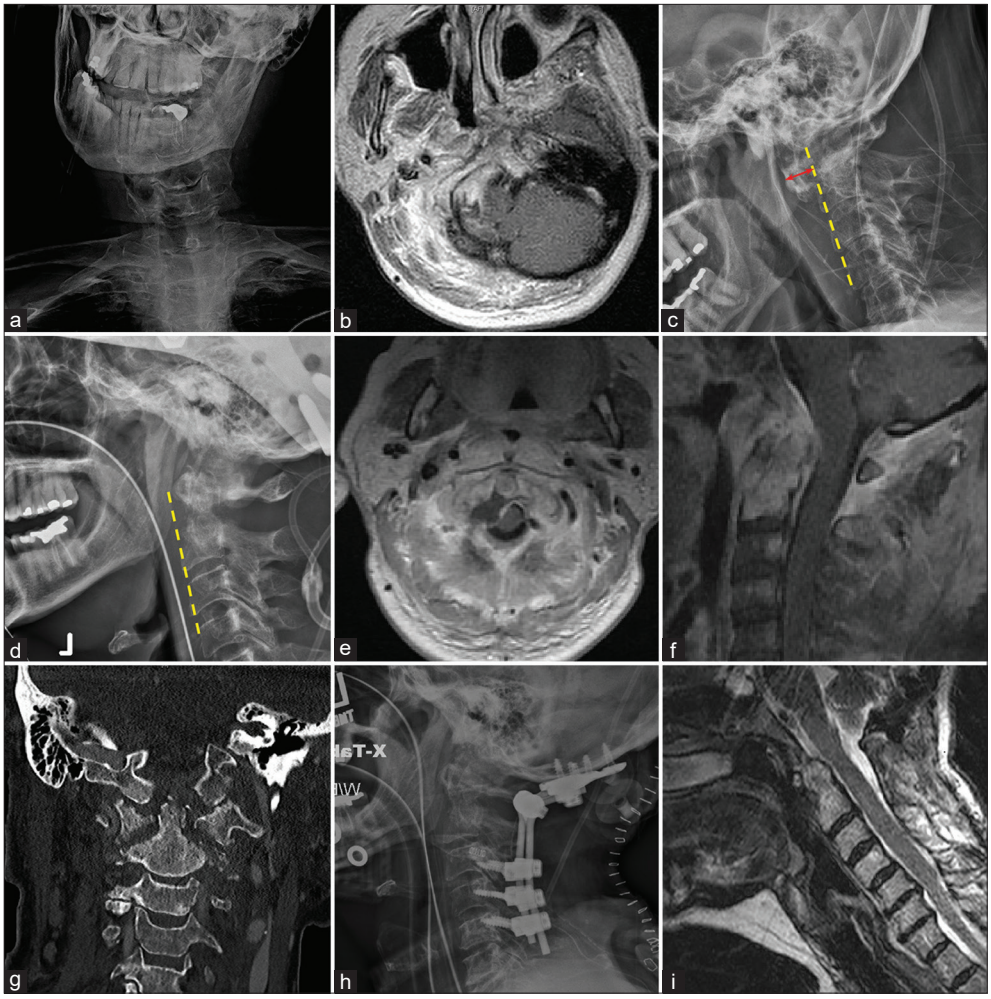

Figure 2: Case 1: X-ray of the neck demonstrating cock-robin position (a); T1 magnetic resonance imaging (MRI) of suboptimal quality due to neck tilt (b); lateral X-ray of the craniocervical junction showing increased atlanto-dental interval marked by the red arrow (c); post traction imaging demonstrating complete reduction on lateral X-ray (d) with extensive inflammation seen on axial (e) and sagittal (f) T1 MRI and coronal computed tomography (g) of the cervical spine; postoperative lateral neck X-ray (h); and sagittal T2 MRI (i) demonstrating proper instrumentation and alignment. Yellow dashed line indicates anterior alignment of the cervical spine.

and progressive dysphonia, dysphagia, and quadriparesis (1/5 upper and 3/5 lower extremities with high cervical pin level). On examination, his neck was locked in the "cock-robin" position [Figure 3a]. His WBC was elevated at 22,000/mm^3, and the cervical CT showed AARS with destruction of the dens and bilateral facet joints [Figures 3b-d]. The cervical MRI showed retropharyngeal enhancement and C1 cord compression, while X-rays revealed subluxation and an increased atlantodental interval [ADI, Figures 3e-f]. Following the application of 15-lbs cervical traction, his motor exam improved to 4/5 in the upper and 5/5 in the lower extremities; traction was then converted to a halo device. The cervical CT confirmed improved alignment with reduction of AARS and resolution of cord compression [Figures 3g-h]. He subsequently underwent a posterior occiput-C5 fusion and was braced in a rigid collar [Figure 3i]. The blood cultures grew methicillin-susceptible *staphylococcus aureus*, and he was managed with appropriate antibiotic therapy. Although his hospital course was uncomplicated, he was lost to follow-up after discharge.

DISCUSSION

Nontraumatic AARS in adults is usually attributable to infection [Table 1].[1-3,5-8,10] Patients who fall into Types I and II AARS according to the Fielding and Hawkins classification [Figure 1] may be classically managed with bracing, while Type III and IV (i.e., posterior subluxation) in adults usually warrant surgical reduction with instrumented fusions.[4]

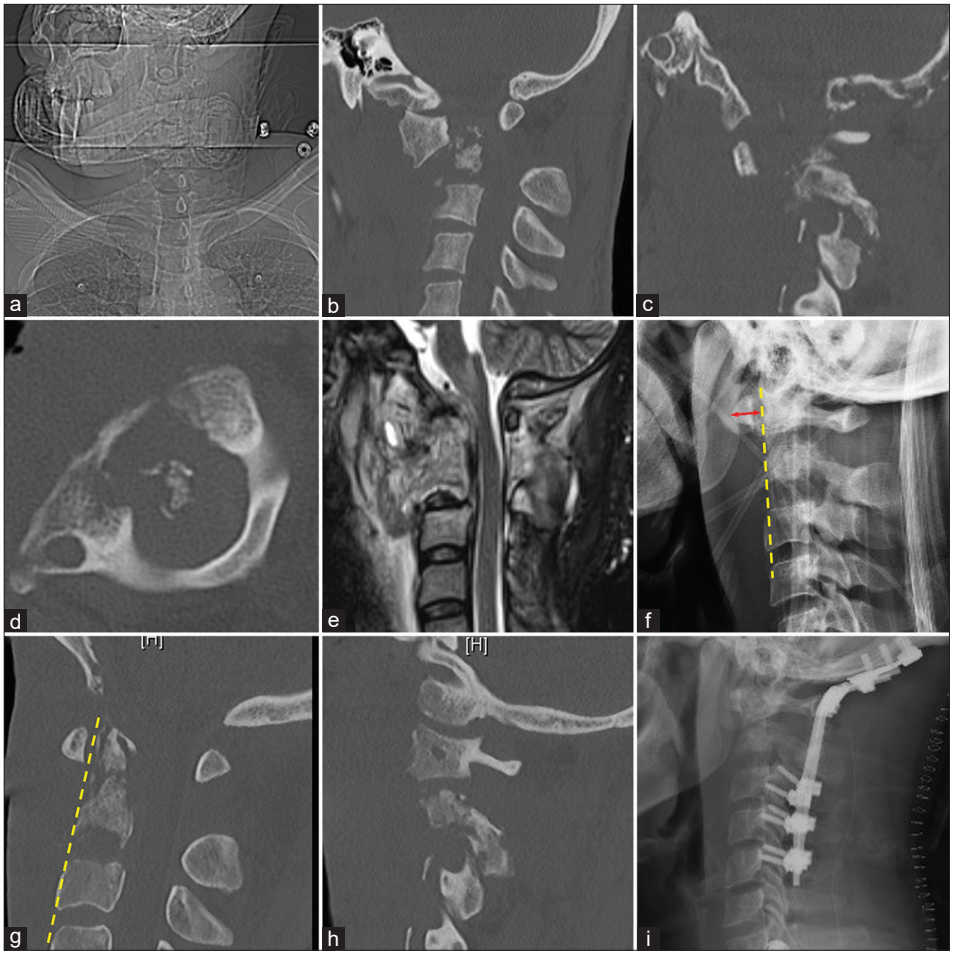

Figure 3: Case 2: X-ray of the neck showing cock-robin position (a); computed tomography (CT) of the cervical spine demonstrating anterior C1 subluxation with destruction of the odontoid process (sagittal, b) and facet joint (coronal, c) with rotation seen in the axial plane (d); sagittal T2 magnetic resonance imaging demonstrating inflammatory process involving C1-2 with cord signal change (e); lateral neck X-ray with increased atlanto-dental interval marked by the red arrow (f); post-traction CT showing restoration of alignment at midline (g) and facet joint (h); and postoperative lateral X-ray with proper instrumentation (i). Yellow dashed line indicates anterior alignment of the cervical spine.

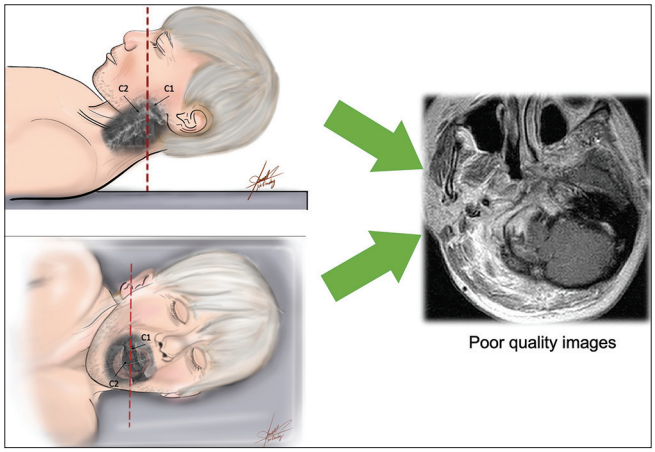

Figure 4: Illustration demonstrating how suboptimal imaging is produced in the setting of severe neck angulation.

Utility of CT and MR studies for diagnosing AARS

Patients with suspected nontraumatic AARS should undergo CT and MRI to confirm the diagnosis but only following adequate reduction, as placing them flat in the scanner before stabilization is not safe [Figure 4].

Protocol for diagnosing AARS

The diagnosis of AARS should be established based on clinical evaluation and X-ray studies. Advanced imaging, including CT and MRI, should be delayed until adequate traction (i.e., 5-10-15 lbs) or halo stabilization has been achieved. Surgical instrumentation/fusion in adults may then be warranted based on whether patients fulfill Type III or IV Fielding and Hawkins classification criteria.

CONCLUSION

Nontraumatic AARS is rare in adults. We support a stepwise management strategy that first includes traction for C1/C2 reduction and halo stabilization, followed by advanced MR/CT imaging to document cord decompression and determine whether fusion is warranted.

Declaration of patient consent

Patients' consent not required as patients' identities were not disclosed or compromised.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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