

Chronic neglected irreducible atlantoaxial rotatory subluxation in adolescence

ABSTRACT

Atlantoaxial rotatory fixation (AARF) is a rare condition and delayed diagnosis. We report a case of chronic neglected atlantoaxial rotatory subluxation in adolescence child that was treated by serial skull traction followed by posterior fusing by method pioneered by Goel *et al*. A 15-year-old male presented with signs of high cervical myelopathy 2 years after trauma to neck childhood. There was upper cervical kyphosis, direct tenderness over C2 spinous process, atrophy of both hand muscles with weakness in grip strength. Reflexes in upper and lower extremities were exaggerated. Imaging showed Type 3 (Fielding and Hawkins) rotatory atlantoaxial dislocation (AAD). Treatment options available were 1. Staged anterior Transoral release & reduction followed by posterior fusion described by Govender and Kumar *et al*, 2. Posterior open reduction of joint and fusion, 3. Occipitocervical fusion with decompression. Our case was AARF presented to us with almost 2-year post injury. Considering complications associated with anterior surgery and posterior open reduction, we have opted for closed reduction by serially applying weight to skull traction under closed neurological monitoring. We have serially increased weight up to 15 kg over a period of 1 week before. We have achieved some reduction which was confirmed by traction lateral radiographs and computerized axial tomography scan. Residual subluxation corrected intra-operatively indirectly by using reduction screws in Goel *et al*. procedure. Finally performed for C1-C2 fusion to take care of Instability. We like to emphasis here role of closed reduction even in delayed and neglected cases.

Key words: Atlantoaxial dislocation; atlantoaxial rotatory fixation; rotatory subluxation.

Introduction

Atlantoaxial rotatory fixation (AARF) is a rare condition and often under recognized, resulting in an incorrect or delayed diagnosis.^[1] There is usually a higher incidence in children. The common clinical characteristics are painful torticollis and cock robin position presented with the head tilted to one side and rotated to the other side. Because a clinical diagnosis of AARF is generally difficult, it is often made in the late stage. In some cases, an irreducible or chronic fixation develops.^[2] We report a case of chronic neglected atlantoaxial rotatory subluxation in adolescence child that was treated by serial skull traction followed by posterior fusing by method pioneered by Goel and Laheri.^[3]

Case Report

A 15-year-old male presented with upper cervical kyphosis and signs of high cervical myelopathy almost 2 years after

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initial trauma in childhood. On clinical evaluation, there was upper cervical kyphosis, suboccipital pain, and direct tenderness over C2 spinous process. The neurological evaluation showed atrophy of both hand muscles with weakness in grip strength. Exaggerated reflexes in upper and lower extremities.

Anteroposterior open-mouth radiography [Figure 1] and lateral radiography of the cervical spine [Figure 2] demonstrated rotatory atlantoaxial dislocation (AAD). A computed tomography (CT) scan with reconstruction confirmed the presence of rotatory dislocation [Figures 3 and 4]. The rotation axis was the dens, and the atlas–dens interval was normal. These findings established the diagnosis of rotatory AAD corresponding to Type 3 in the classification scheme developed by Fielding and Hawkins.^[4] Reduction of this 2-year-old dislocation proved difficult and required 10 days of skull traction followed by gentle manipulation skull traction was started immediately at 3 kg and was gradually increased to

15 kg. Cranial nerve function was closely monitored. Although muscle relaxants and analgesics were given, reduction proved difficult, and the possible need for open reduction through the transoral route was discussed with the patient. However, the reduction was obtained after 10 days by manipulation of the neck. The reduction maneuver consisted in increasing the distraction by pulling gently on the neck at the bedside, while the patient was conscious, then in a movement aimed at correcting the lateral flexion and rotation. This movement corrected the torticollis, at the same time producing neck pain. Serial radiological monitoring is done during this period [Figure 5]. Anteroposterior open-mouth radiographs confirmed that the dislocation had been corrected to more acceptable position with some residual subluxation.

To determine whether C2–C1 fusion was required, dynamic radiographs of the cervical spine were obtained [Figure 6]. There was evident instability, in form of recurrent subluxation from reduced position, and the atlas–dens interval



Figure 1: Anteroposterior open-mouth radiography



Figure 2: Lateral radiography of the cervical spine



Figure 3: Computed tomography findings of rotatory dislocation of both atlantoaxial joints. The axis of rotation is the dens

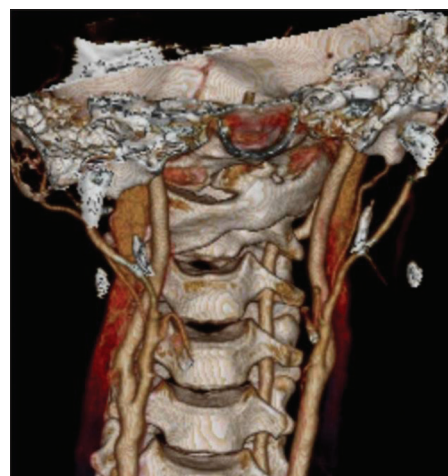


Figure 4: Three-dimensional reconstruction showing rotatory dislocation of both atlantoaxial joints



Figure 5: Serial radiological and anteroposterior open-mouth radiographs confirmed that the dislocation had been corrected to more acceptable position with some residual subluxation

was abnormal (C1–C2, <3 mm). A magnetic resonance imaging (MRI) and reconstruction CT angiogram study were performed to look for posttraumatic spinal cord contusion and, above all, to evaluate the transverse ligament. In addition, the integrity of the medulla, cervical cord, and the vertebral arteries can be demonstrated.^[5-7]

Surgical technique

The patient is positioned prone using a Mayfield head holder (OMI, Inc., Cincinnati, OH, USA). The neck is kept neutral and the head is placed in the military tuck position. The arms are tucked at the sides. The shoulders are retracted caudally using tape. A midline incision is made extending from the suboccipital area to the spinous process of C3. The C2–C3 facet joints are exposed and the dorsal arch of C1 is exposed laterally exposing the vertebral artery in the vertebral groove on the superior aspect of the C1 arch (sulcus arteriosus). The C2 nerve root is identified and is either sacrificed or mobilized inferiorly. Bipolar cautery and hemostatic agents such as gel foam are used to control bleeding from the venous plexus surrounding the C2 nerve root and also surrounding the vertebral artery. The lateral mass of C1 inferior to the C1 arch is exposed after the C2 nerve root has been sacrificed or mobilized inferiorly. The medial wall of the C1 lateral mass is identified using the forward angle curette to palpate the medial limit of screw placement. The medial aspect of the transverse foramen at C1 and C2 can also be identified and serve as a lateral limit for screw placement.



Figure 6: Computed tomography scan showing evident instability in form of residual subluxation from reduced position

Using fluoroscopy, a 3 mm drill bit and guide are used to drill a hole with 10–15° medial angulation to penetrate the anterior cortex of C1. On lateral fluoroscopic imaging, the drill is aimed toward the anterior tubercle of C1 so that the drill penetrates the ventral cortex of the lateral mass midway between the superior and inferior facets of C1. The hole is tapped and subsequently, a C1 lateral mass screw is placed.

We then turn our attention to placing a screw at C2. The C2 pedicle, on the other hand, is the portion of the C2 vertebrae connecting the dorsal elements with the vertebral body (the C2 pedicle is anterior to the C2 pars). The trajectory of the C2 pedicle screw is different than that of C2 pars screw. The entry point for a C2 pedicle screw is in the pars of C2, lateral to the superior margin of the C2 lamina. This point is usually 2 mm superior and 2 mm medial to the entry point for the C2 pars screw that we have just described. Goel's C1 lateral mass screw with C2 pars/pedicle screw fixation. The screw is placed with 15–25° of medial angulations. The thick medial wall of the C2 pedicle will help redirect the screw if necessary and prevent medial wall breakout and entry into the spinal canal. The screw is placed after a drill is used to create the entry hole and after the hole is tapped. The trajectory of the C2 pedicle screw is 20° up angle and 15–25° medial from the entry point. The screws are tightened over a plate or rods are top loaded onto the screw heads. Figure 7: (a) Postoperative lateral X-ray (b) anteroposterior X-ray showing final alignment.

Results

At the 6 months follow-up, he was free from the neck pain and resumed normal activities. These follow-up radiograph images confirmed the atlas and axis have been completely repositioned. The “cockrobin” posture disappeared and fusion was noted after 4–7 weeks with normal alignment of the atlantoaxial joints and the atlantodental interval. Rehabilitation therapy aimed at recovering mobility was started. Twelve months later, the range of motion of the neck was normal in all planes, and a dynamic lateral radiograph

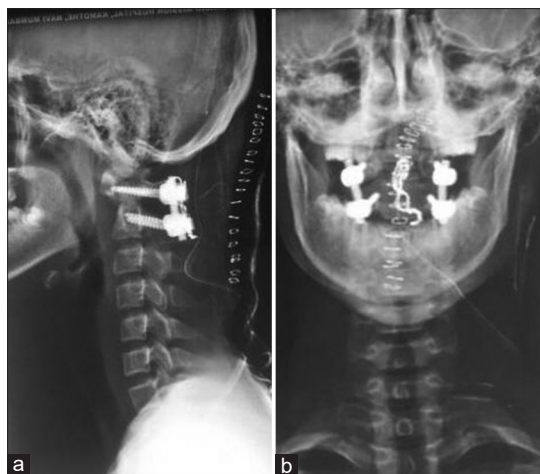


Figure 7: (a) Postoperative lateral X-ray (b) anteroposterior X-ray

of the cervical spine confirmed that the atlantoaxial joint was stable.

Discussion

Rotatory dislocation is an abnormality seen in the upper cervical spine, where most of the cervical rotation occurs. Rotatory AAD was first reported in children and is uncommon in adults, most case being seen after severe trauma.^[8] Few cases of rotatory AAD have been reported in adults perhaps because the cause may often be severe trauma responsible for lethal injuries.^[9,10]

Factors that influence therapeutic choices include time since injury, ease of reduction, presence of neurological compromise, and stability of the lesion. As in all dislocations, the treatment consists in immediate reduction. If the trauma was minor, a collar, cervical traction,^[11] or manipulation may allow reduction of the dislocation to be obtained. However, the success rate of conservative treatment drops if the diagnostic delay exceeds 1 month.^[12] When the diagnosis is delayed, reduction of the dislocated atlantoaxial joint can be extremely difficult, requiring several weeks of skull traction without over distraction in unstable lesions.^[10]

Although the natural history of the untreated condition is not known, Fielding and Hawkins^[4] reported a follow-up for 8 years of two patients who refused surgery. Spontaneous correction of the clinical deformity occurred in one and in the other the deformity was unchanged. High success rates have been reported with conservative therapy in patients with promptly diagnosed, stable, and rotatory AAD^[13,14] not associated with transverse ligament disruption.

The transverse ligament is a primary stabilizing component of the atlantoaxial joint. On lateral radiographs of the cervical

spine in flexion and extension, an atlas–dens interval (i.e., the distance between the anterior arch of C1 more than 3 mm is considered unstable. However, the choice between conservative therapy and fusion depends on whether or not the lesion is stable. The transverse ligament is intact in patients with bilateral rotator AAD but is ruptured in unilateral dislocation.^[15] In patients with instability due to transverse ligament disruption, C2–C1 fusion by screws is the option associated with the smallest degree of motion range limitation.^[16]

In chronic AARF, it is essential not to reduce the joint after the transoral release. Fang and Ong^[17] reported a case of chronic AAD in which a blunt hook was used to reduce the joint. Hemorrhage occurred on several occasions and despite adequate resuscitation, the patient developed a fever and died 9 days after transoral surgery. A tear of the vertebral artery and vein was revealed at postmortem.

Govender and Kumar concluded that although AARF is uncommon, early recognition and treatment may prevent subsequent instability and deformity. MRI of the atlantoaxial complex and MRA of the vertebral arteries are essential before surgery in patients with chronic AARF. A staged procedure was safe and useful in correcting the clinical and radiological deformity in chronic AARF after trauma.^[18]

Reduction is difficult if the diagnosis is delayed by more than 1 month. Skull traction is indicated, if necessary in association with direct manipulation of the neck.^[19]

Conclusion

Rotatory AAD is relatively rare in adults but can be caused by neck injuries; diagnosis is easily missed if the neurological evaluation is normal. An anteroposterior open-mouth plain radiograph suggests the diagnosis, which can be confirmed by CT. Reduction is difficult if the diagnosis is delayed by more than 1 month.

Skull traction is indicated, if necessary in association with direct manipulation of the neck. Stability is dependent on whether the transverse ligament is intact and can be evaluated by measuring the atlas–dens interval on dynamic cervical radiographs and the atlas–axis angle on CT scans in maximum rotation. When the atlantodental exceeds 5 mm, there is instability of the C1–C2 complex and posterior C1/2 fixation is indicated.

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Conflicts of interest

There are no conflicts of interest.

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