Case Report

Rotatory atlantoaxial dislocation presenting as spinal kyphoscoliosis

ABSTRACT

A 16-year-old male presented with primary complaint of worsening dorsal spinal kyphoscoliosis (SKS) for 3 years. More recently, he developed spasticity in legs, breathlessness on mild exertion, and sleep apneas. Apart from SKS, investigations revealed rotatory atlantoaxial dislocation. Atlantoaxial fixation resulted in rapid recovery from all symptoms including from spinal deformity. Observations in this patient suggest that rotatory dislocation can be a cause of spinal deformity.

Keywords: Atlantoaxial dislocation, kyphoscoliosis, syringomyelia

INTRODUCTION

We report a case where spinal kyphoscoliosis (SKS) was associated with rotatory atlantoaxial dislocation. Such an association has never been recorded. The cause-effect relation of rotatory atlantoaxial dislocation and SKS and treatment strategy is discussed.

CASE REPORT

A 16-year-old boy presented with progressively increasing deformity of the chest wall and spine for the past 3 years. He was unable to sleep supine on his back and preferred lateral position for 3 years. For about 6 months, he had pain in the nape of neck and noticed that he was not able to run as fast as his classmates. He also had tingling paresthesia and weakness of the right hand that lead to difficulty performing fine tasks and holding heavy objects. He complained of breathlessness on exertion. On further enquiry, he admitted to having difficulty in breathing at night; sleep apnea's leading to sudden awakening. Neurological examination revealed spasticity in all 4 limbs. There was mild (Grade 4/5) weakness in the right handgrip. There was spasm of the muscles of nape of neck. There was no neck deformity. His Goel clinical grade was Grade 2.[1] Investigations of the spine and chest revealed dorsal kyphoscoliosis. His preoperative scoliosis curve measured using the Cobb's angle was 60° and his thoracic kyphosis was 40°. Dynamic computed tomography of the cervical spine did not reveal any abnormal alteration in the atlantodental interval or any evidence of neural compression. However, there was evidence of rotatory atlantoaxial dislocation[2,3] [Figure 1]. The patient underwent atlantoaxial fixation using lateral mass plate and screw technique as described by us earlier.[4,5] During surgery, the atlantoaxial facetal articulation was identified to be remarkably unstable. The articular cavity was widely opened, articular cartilage was denuded, facets were realigned using the technique of manual manipulation described earlier and subsequent instrumentation was done. The muscles attached to the spinous process of axis were sharply cut, bones of the posterior elements of axis and atlas were decorticated to make them suitable host for bone graft. Bone graft was harvested from iliac crest.

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Submitted: 16-Jan-21 **Accepted:** 17-Jan-21
Published: 04-Mar-21

Access this article online	
Website: www.jcvjs.com	Quick Response Code
DOI: 10.4103/jcvjs.JCVJS_6_21	

How to cite this article: Goel A, Vutha R, Shah A, Rai SK. Rotatory atlantoaxial dislocation presenting as spinal kyphoscoliosis. J Craniovert Jun Spine 2021;12:99-101.

The patient improved in his clinical features in the immediate postoperative period. Relatives noticed improved volume of voice. He could sleep in supine position in the postoperative period, a position that he had not adopted for months. He also improved in all his limb symptoms, could walk better and the power in the right hand grip recovered. At a follow-up of 18 months, there was improvement in his SKS. His neurological examination was normal and he had joined back school and was performing all his normal activities. Postoperative investigations revealed atlantoaxial arthrodesis. Postoperative X-ray of chest and dorsal spine revealed improvement in his SKS. The coronal Cobb's improved to 50° and the thoracic kyphosis improved to 38° [Figure 1].

DISCUSSION

While acute atlantoaxial instability is associated with acute clinical symptoms, chronic or long-standing atlantoaxial dislocation is generally associated with chronic or longstanding neural and musculoskeletal changes that "frequently" include SKS.[6] Musculoskeletal alterations include short neck, torticollis, Klippel–Feil abnormalities, and platybasia and neural alterations include Chiari formation and syringomyelia.[1,6,7] It was identified that these musculoskeletal and neural alterations were natural designs aimed to protect the body and to counter potential or manifest atlantoaxial instability.[7] The more crucial issue was the identification of the fact that such musculoskeletal abnormalities are potentially "reversible" following atlantoaxial stabilization. For the first time in the literature, recovery from otherwise considered "fixed" spinal abnormality of torticollis and improvement in the length of the neck and recovery from SKS following atlantoaxial fixation was reported.[2,3,6,7]

More recently, we identified that in cases with basilar invagination and chronic atlantoaxial instability, short neck may even be associated with short head and short spine.[8-10] It appears that like the more commonly identified "short neck" results in torticollis, SKS may be a manifestation of "short spine." Apart from cosmetically negative issue of SKS, other symptoms such as breathing difficulty, voice alteration, pain in the back and shoulders, and spasticity in the legs are generally attributed to the spinal bony and neural deformation associated with SKS and probable compressive and deforming effects on the spinal cord and lungs. Our recent studies identified that chronic atlantoaxial instability, more often of central or axial type, when associated with or without Chiari formation, with or without syringomyelia and with or without bone malformations at the craniovertebral junction can present with major symptoms of voice volume and quality alterations and breathing disturbances apart from motor and sensory dysfunction.[11] The identification of the fact that atlantoaxial instability can be nodal point of pathogenesis of the entire symptom complex and

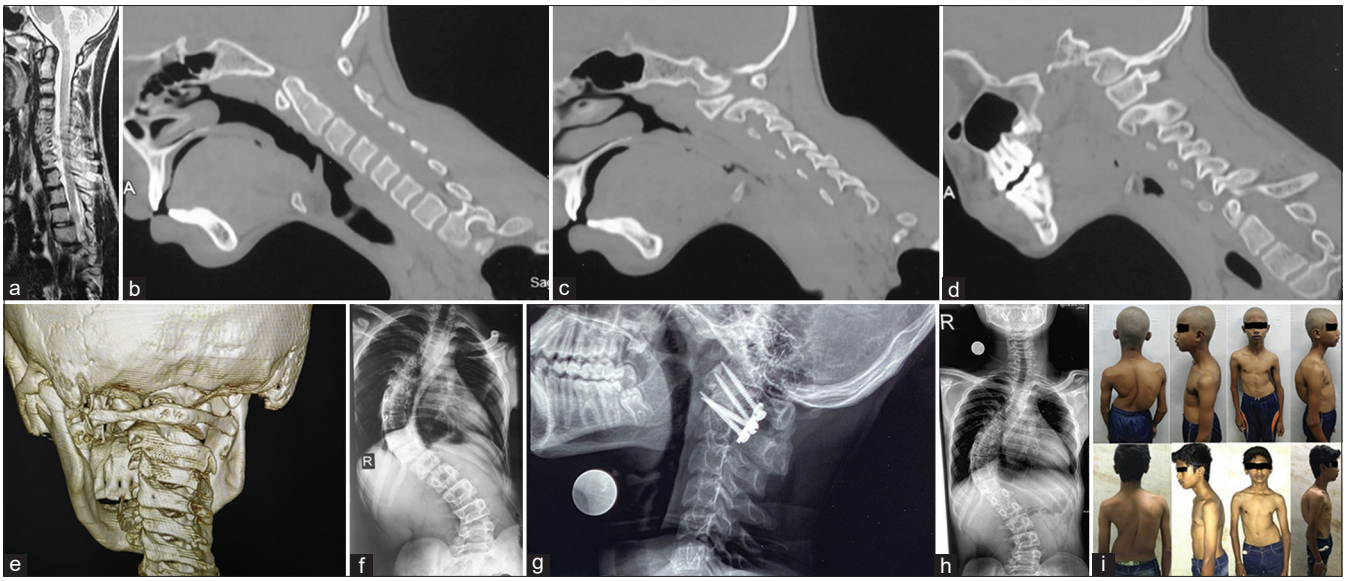

Figure 1: Images of a 16-year-old male patient. (a) T2-weighted magnetic resonance imaging showing no evidence of compression of the neural structures at craniovertebral junction. (b) Computed tomography scan with the neck in flexion showing no evidence of atlantoaxial instability. (c) Computed tomography scan with the head in flexion showing the right sided facets. The facet of atlas is dislocated anterior to the facet of axis. (d) Computed tomography scan with the head in flexion showing the left sided facets. The facet of atlas is dislocated posterior to the facet of axis. (e) Three-dimensional computed tomography scan image showing rotatory atlantoaxial dislocation. (f) Anteroposterior view of the X-ray chest showing spinal kyphoscoliosis. (g) Lateral radiograph showing atlantoaxial fixation and fusion. (h) Postoperative plain radiograph showing reduction of spinal kyphoscoliosis. (i) Images of the patient (preoperative-upper line) and postoperative (lower line)

musculoskeletal disability that includes SKS can lead to directing the treatment to atlantoaxial stabilization.

Rotatory atlantoaxial dislocation is usually identified in young children following minor or insignificant trauma. The classically described "cock-robin" head posturing is a frequent observation. Apart from neck deformity, the patient more often has no pain or neurological symptom. In the year 2011, we described the technique of facetal manipulation, realignment, and fixation as treatment for rotatory dislocation.[2] Considering the difficulty in surgical treatment and potential for neurological complications in an otherwise intact patient, conservative treatment option has been generally preferred. Long-term effect of neglected, untreated, or unrecognized rotatory atlantoaxial dislocation has only infrequently been described.[12] While the progression of rotation and or torticollis and mal-aligned positioning of the neck have been described, its effect on the rest of the spine has not been discussed.

The presented patient had rotatory dislocation, but there was no apparent torticollis. It seems that SKS was a natural spinal maneuvering to compensate for the torticollis and permit looking straight ahead. Atlantoaxial stabilization resulted in rapid recovery in all neurological symptoms and of SKS. It does suggest the primary nodal point of pathogenesis of the structural abnormality and functional disability in the presented case was neglected or unrecognized rotatory atlantoaxial dislocation. It is true that the extent of recovery in SKS was not as dramatic as reported by several authors who resort to multilevel decompression and stabilization. However, avoiding a "major" fixation surgical procedure and retaining suppleness and movements of the spine was a major advantage.

CONCLUSION

Chronic rotatory atlantoaxial dislocation can be a cause of SKS. The treatment in such cases should be directed toward atlantoaxial stabilization.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Goel A. Is atlantoaxial instability the cause of chiari malformation? Outcome analysis of 65 patients treated by atlantoaxial fixation. J Neurosurg Spine 2015;22:116-27.
2. Goel A, Shah A. Atlantoaxial facet locking: Treatment by facet manipulation and fixation. Experience in 14 cases. J Neurosurg Spine 2011;14:3-9.
3. Goel A. Torticollis and rotatory atlantoaxial dislocation: A clinical review. J Craniovertebr Junction Spine 2019;10:77-87.
4. Goel A, Desai K, Muzumdar D. Atlantoaxial fixation using plate and screw method: A report of 160 treated patients. Neurosurgery 2002;51:1351-7.
5. Goel A, Laheri VK. Plate and screw fixation for atlanto-axial dislocation. (Technical report). Acta Neurochir (Wien) 1994;129:47-53.
6. Goel A, Vutha R, Shah A, Dharurkar P, Jadhav N, Jadhav D. Spinal kyphoscoliosis associated with chiari formation and syringomyelia 'recovery' following atlantoaxial fixation: A preliminary report and early results based on experience with 11 surgically treated cases. World Neurosurg 2019;125:e937-e946.
7. Goel A, Shah A. Reversal of longstanding musculoskeletal changes in basilar invagination after surgical decompression and stabilization. J Neurosurg Spine 2009;10:220-7.
8. Goel A. Short neck, short head, short spine, and short body height – Hallmarks of basilar invagination. J Craniovertebr Junction Spine 2017;8:165-7.
9. Goel A, Jain S, Shah A. Radiological evaluation of 510 cases of basilar invagination with evidence of atlantoaxial instability (Group A basilar invagination). World Neurosurg 2018;110:533-43.
10. Goel A, Nadkarni T, Shah A, Sathe P, Patil M. Radiological evaluation of basilar invagination without obvious atlantoaxial instability (Group B-basilar invagination): An analysis based on a study of 75 patients. World Neurosurg 2016;95:375-82.
11. Goel A, Prasad A, Shah A, Gore S, Dharurkar P. Voice quality affection as a symptom of chiari formation. World Neurosurg 2019;121:e296-e301.
12. Goel A, Jain S, Shah A. Management of a case of neglected atlantoaxial rotatory dislocation. Neurol India 2017;65:1170-3.