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Atlantoaxial instability associated with pan cervical vertebral fusion: Report on management of 4 cases

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Abstract

We report a series of four patients aged 4, 5, 14, and 27 years (1 male and 3 female patients) with severe shortening of the neck and torticollis since early childhood who presented with complaint of pain in the nape of neck as the primary symptom. All four patients had relatively well preserved neurological functions. One patient had vertical mobile and reducible atlantoaxial dislocation, and 3 patients had anteroposterior mobile and reducible dislocation. There was assimilation of atlas in 1 patient. The arch of atlas was bifid in 3 patients. Two patients underwent atlantoaxial fixation. Both the patients were relieved of neck pain after their surgery. The potential surgical difficulties due to the presence of severe shortening of neck height and marginal presenting symptoms favored conservative observation in the other 2 patients. Follow-up ranged from 6 to 84 months. All patients are functionally and socially active.

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Full Text

Atlantoaxial instability has been identified to be frequently associated with assimilation of atlas, C2-3 fusion, and Klippel–Feil abnormality.[1],[2] We report four cases wherein atlantoaxial instability was associated with pancervical vertebral body fusion. Two cases underwent atlantoaxial fixation. Severely short neck resulted in difficulty in exposure of the atlantoaxial joint and fixation. Our literature search did not reveal reports of similar cases.

Case Spectrum

From 2009 to 2016, we identified 4 patients (1 male and 3 female patients aged 4, 5, 14, and 27 years, respectively)

having pancervical vertebral fusion that was associated with mobile atlantoaxial instability. All 4 patients presented with severely short neck and restricted neck movements. The primary complaint at the time of presentation in all the 4 patients was pain in the nape of neck that worsened on neck movements. One patient also had occasional episodes of dyspnea. She was not greatly discomforted by this symptom. On neurological examination, there was hyperreflexia in 2 patients. Apart from this, there were no neurological deficits. Investigations included dynamic computed tomography (CT) scan and magnetic resonance imaging (MRI) in all the 4 patients [Figure 1] and [Figure 2]. In one patient, additionally, a three-dimensional (3D) printed model was made.[3] All the patients had complete fusion of the subaxial spine extending up to the C7 level in 3 patients and the C6 level in 1 patient. The imaging gave an impression of a "bamboo spine," as seen in ankylosing spondylitis. However, all the 4 patients were HLA B27 negative. Investigations showed assimilation of atlas in 1 patient and bifid anterior and posterior arches of the atlas in 3 patients. The posterior bifid was uniformly large and made a huge space for the dura posteriorly. In addition, 1 patient had a wide posterior bifid C2 laminar arch. The bifid processes gave the appearance of a "natural laminectomy." MRI showed the presence of a large subarachnoid space, which we have referred to earlier as external syrinx.[4] Three patients had an anteroposterior mobile atlantoaxial dislocation [Figure 1], and 1 patient had a vertically mobile atlantoaxial dislocation [Figure 2]. In 1 patient, there was Group B basilar invagination. In view of the marginal degree of complaints, anticipation of surgical difficulties, and nonacceptance of the potential surgical risks by the patient and the relatives, 2 patients did not undergo surgery and are under clinical observation. The other two patients underwent surgery. The patients were operated in the prone position with the head in a "floating" position, under Gardner Well's cervical traction, as discussed by us earlier.[5],[6] After the initial subperiosteal dissection, the lamina of the C2 vertebra was identified. The C1-2 joint was exposed in both the patients with considerable difficulty and significant venous bleeding. The joints were markedly unstable in both cases. The facet of atlas was laterally positioned and had an oblique profile.[7] The two halves of the arches of atlas were markedly mobile, even on mild touch. The articular surfaces were widely denuded of the cartilage present on them, and the articular cavity was packed with bone graft pieces harvested from the iliac crest. Goel's C1 lateral mass and C2 pedicle screw fixation with plate and screw was subsequently performed.[5],[6] Bone graft harvested from the iliac crest was then placed over the decorticated bone over the lamina of the axis and the part of the posterior arch of atlas lateral to the bifid region. Both the patients showed improvement in their neck pain following the surgery. The patients were advised to wear a Philadelphia collar for 2 months after the surgery, and neck movements were restricted during the time. At an average follow-up of 18 months, both the operated patients are well and symptom free. The two non-operated patients continue to be under observation. During the period of follow-up, both patients did not show worsened symptoms or neurological deficits sufficient to force them to undertake the surgical option. {Figure 1}{Figure 2}

Discussion

Cervical vertebral body fusions are relatively uncommon but have been recorded and frequently reported. They have been often associated with basilar invagination and atlantoaxial instability.[1],[8],[9] Bone fusions generally occur in consort with a short neck and torticollis. Failure of segmentation and embryonic dysgenesis has been classically implicated as a primary cause of this morphologic abnormality.[10] However, the question is whether short neck results in bone fusions or if bone fusions result in short neck. In other words, it is unclear if chronic and longstanding atlantoaxial instability is the primary event and bone fusions are a result of chronic need for addressing the ensuing muscle spasm and shortening of neck. It is also debated if bone fusions are a form of natural protection due to atlantoaxial instability or it is a part of a pathological cohort. Another relevant point of discussion is whether or not extensive fusion of the subaxial cervical spine causes atlantoaxial instability just like adjacent segment degeneration occurs in the subaxial cervical spine after fusion that may lead to bony ankylosis later on. In all our cases, there was no fusion abnormality of the dorsal or lumbar spines and neither was there any suggestion of generalized ossification of the anterior or posterior longitudinal ligaments.

Bone fusions are more often located above and/or below the site of maximum neural compression at the tip of the odontoid process and are identified as assimilation of atlas and C2-3 vertebral fusions.[1] Less frequently, bone fusions occur in subaxial bones and in lower cervical spine, and such fusions are labeled as Klippel–Feil abnormality.[2],[11] Platybasia and reduction in the size of clivus are also frequent associations.

In 2009, we identified that atlantoaxial instability is the primary pathology and observed that bone fusions are a secondary and probably a protective natural response.[1] We speculated that longstanding neck muscle spasms and related muscle contractures and restricted neck movements are probably the incriminating issues that first result in reduction in the disc space height, secondary osteophyte formation, and subsequently bone fusions.[1] Other musculoskeletal features include hyperextension of the neck and restriction of neck flexion. We have recently identified that even neural malformations such as Chiari 1 malformation and syringomyelia are secondary formations and are a consequence of subtle and chronic atlantoaxial instability.[12],[13] Identification of the fact that several musculoskeletal and neural alterations are reversible following atlantoaxial stabilization provides credibility to the hypothesis. Our study implicates focal and generalized spinal instability as the cause of osteophyte formation, retro-odontoid ossification/calcification, ossification of posterior longitudinal ligament, and bone fusion. Accordingly, we have proposed that "only fixation" can form the basis of treatment of degenerative spine, ossified posterior longitudinal ligament, and basilar invagination.[14],[15],[16] Although never clinically observed, even in the cases reported, we speculated that there is a potential for regression of osteophytes and for reversal of bone fusions following atlantoaxial fixation. We reported regression of the retroodontoid "pseudotumor" and "pannus" following atlantoaxial fixation.[17],[18],[19]

Atlantoaxial joint is the most mobile joint of the neck. To facilitate circumferential movement, the joint architecture is unique wherein the articular surfaces are round and flat. While this structural formation facilitates unrestricted movements, the joint is most prone to develop instability. Instability at the atlantoaxial joint has been traditionally diagnosed by an abnormal increase in the atlantodental interval on dynamic neck flexion-extension images. We have recently identified that the atlantoaxial instability can be vertical,[20] lateral,[7],[21],[22] circumferential, axial, or central in nature.[23] We classified atlantoaxial instability on the basis of facetal malalignment.[21] Essentially, atlantoaxial instability can be subtle, chronic, or longstanding in nature, and cord compression may not be an early or a prominent feature. In such cases, the neurological myelopathy related symptoms are absent or subtle, and longstanding and secondary musculoskeletal and neural malformations form prominent associations. The neural soft tissue and bone alterations assist in delaying or stalling the neurological sequel of instability. All our patients had only marginal symptoms despite the presence of several and severe bone and soft tissue abnormalities. Atlantoaxial instability is often associated with basilar invagination, Chiari 1 malformation, syringomyelia, degenerative spinal changes, and ossified posterior longitudinal ligament, among other abnormalities.[4],[11],[24],[25],[26] Identification of the fact that atlantoaxial instability can be present despite the absence of abnormality in the atlantodental interval has certainly expanded the scope of understanding of this subject. One of our patients had vertical mobile and reducible atlantoaxial instability. Such instability is a result of incompetence of facets and laxity of ligaments. In 3 patients (cases 2,3, and 4), there was additional presence of bifid posterior arch of atlas. Our fixation procedure involved lateral mass fixation on each side. However, considering that there is a potential for two fixed segments on each side to move relative to each other in a horizontal perspective, a cross clamp fixation may possibly be the option.[7] However, such a procedure was not adopted. All 4 cases had relatively well-preserved neurological state despite evidence of instability at the atlantoaxial joint, marked shortening of the neck and torticollis. Moreover, envisaging the potential difficulties in exposure of the atlantoaxial joint due to severe shortening of the neck, surgery was avoided in cases 1 and 2, despite the presence of neck pain as a significant symptom in both patients and episodic dyspnea in 1 patient. However, relatively significant neck pain and torticollis forced the other 2 patients to undergo surgical treatment. The atlantoaxial joint was identified to be markedly unstable in these 2 cases.

Conclusion

Chronic atlantoaxial instability can be associated with pancervical spinal fusion. Atlantoaxial fixation is the mode of treatment.

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.

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Conflicts of interest

There are no conflicts of interest.

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