Anterior atlas fracture following suboccipital decompression for Chiari I malformation

Report of two cases

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Chiari I malformation, a congenital disorder involving downward displacement of the cerebellar tonsils through the foramen magnum, is often treated surgically by performing suboccipital craniectomy and C-1 laminectomy. The authors report two cases in which fracture of the anterior atlantal arch occurred during the postoperative period following Chiari I decompression and C-1 laminectomy causing significant neck pain. The findings indicate that interruption of the integrity of the posterior arch of C-1, iatrogenically or otherwise, confers increased risk of anterior arch fracture. A C-1 fracture should therefore be considered in the differential diagnosis of posterior cervical pain in patients who have previously undergone decompression for Chiari I malformation.

KEY WORDS • atlas fracture • Chiari malformation • cervical instability

Fractures of the first cervical vertebra have been estimated to account for 1 to 3% of all spinal column injuries. Atlas fractures, frequently caused by motor vehicle accidents and traumatic falls, seldom result in neurological injury in patients who survive to reach medical attention, largely because of the capacious canal diameter that is characteristic of this spinal level. In addition, centripetal splaying of the C-1 bone ring resulting from an axial loading force, the underlying mechanism of injury, also contributes to the rarity of neurological compromise following this insult. Diagnosis is confirmed rather easily based on either plain radiography or CT studies, and treatment is applied through various approaches in which stabilization of the upper cervical spine during bone healing is the principle goal, that is, with either an external orthosis or an instrumented internal fixation.

Although not yet described in the literature, it is our contention that patients who have undergone a C-1 laminectomy have an increased risk of developing an anterior atlas fracture. Although the underlying mechanism of this fracture has yet to be clearly identified in this context, iatrogenic disruption of the posterior arch and its associated musculoligamentous structures, both important supportive elements of the craniovertebral junction, might confer an increased propensity for fracture of the anterior C-1 arch. In this report, we present two case reports of patients who sustained atlas fractures following suboccipital craniectomy and C-1 laminectomy in the decompressive treatment of Chiari I malformation.

Case Reports

Case 1

Presentation and Examination. This 18-year-old woman whose medical history was significant for mitral valve prolapse initially presented in 1999 with right-sided torso and upper-extremity numbness and tingling, which she had been experiencing for 1 year. She also suffered from intermittent occipital headaches, although these were partially relieved by oral analgesic agents. Results of neurological examination conducted at this time were remarkable only for diminished sensation to light touch along the right torso and extending inferiorly to the hip. Magnetic resonance imaging studies revealed a Chiari I malformation in association with a diffusely loculated cervicothoracic spinal syrinx (Fig. 1 upper left). The cerebellar tonsils were displaced downward to slightly above the posterior arch of C-1.

Operation. Shortly after diagnosis was made, the patient underwent suboccipital craniectomy together with C-1 laminectomy and placement of a dural patch graft. She experienced excellent improvement of her preoperative symptoms, and resolution of the syrinx was radiographically documented (Fig. 1 upper center).
Postoperative Course. Approximately 5 months following surgery, the patient returned to our service with progressive posterior cervical pain that had begun following a violent coughing episode. Plain radiographs demonstrated a nondisplaced linear fracture involving the anterior arch of C-1. Initially, she was treated conservatively by using a hard cervical collar and oral narcotic agents. After 3 months of inadequately treated cervical pain, the patient underwent repeated neuroimaging studies, which revealed 4 mm of displacement from the fracture line (Fig. 1 upper right). Given her unabated pain and the presence of upper cervical instability, internal surgical fixation was performed. To this end, the patient underwent posterior spinal fusion involving the placement of C1–2 transarticular screws and lateral mass plates from C-2 to C-3, C1–2 lateral sublaminar wires, and an autogenous iliac crest bone graft (Fig. 1 lower right). The procedure was performed without any complication, and the patient experienced significant relief of her preoperative cervical pain less than 2 weeks after the procedure. Moreover, follow-up CT studies of the cervical spine revealed a healed atlantal fracture 6 months postoperatively, and the patient experienced no further pain.

Case 2

Presentation and Examination. This 49-year-old woman with a medical history significant for hypertension, diabetes mellitus, and chronic obstructive pulmonary disease initially presented in 2002 with a symptom complex that included occipital headache in conjunction with numbness and tingling affecting the right side of her face as well as her arms and legs bilaterally. She also had difficulty with swallowing as well as nausea and vomiting. Results of neurological examination at this time were remarkable only for a patchy decrease in sensation to light touch in the distribution of her subjective numbness and tingling. Magnetic resonance imaging studies revealed a Chiari I malformation in the absence of a cervical syrinx (Fig. 2 left).

Operation. In an effort to alleviate her symptoms and reduce the likelihood of future complications, the patient underwent suboccipital craniectomy with C-1 laminectomy and duraplasty. She experienced excellent resolution of her symptoms and no immediate postoperative complication.

Postoperative Course. Approximately 6 months following Chiari I decompression, the patient began to experience severe posterior cervical pain that could not be historically linked to any inciting event. Flexion–extension cervical spine radiographs revealed no overt spinal instability. A fracture involving the anterior arch of C-1, which had not been clearly visible on plain x-ray films (Fig. 2 upper right) was demonstrated on CT studies. The patient was initially treated conservatively for 3 months with the aid of a hard cervical collar. Given the persistent cervical
pain and the absence of fusion, primary surgical intervention was considered appropriate. Nonetheless, because of the rotational limitations imposed by a C1–2 fusion, the anatomical course of her vertebral arteries, and the small lateral masses, an alternative therapy to instrumented surgical fusion was performed. To this end, fusion of the anterior C-1 arch fracture was performed using bone morphogenetic protein–2 through a standard anterior cervical incision, followed by placement of an orthotic halo for 3 months. Shortly after the procedure, she experienced significant relief in posterior cervical pain and an evolving fusion was radiographically verified on follow-up neuroimaging less than 3-months postoperatively (Fig. 2 lower right).

Discussion

The Chiari I malformation, initially introduced in the monograph published by John Cleland in 1883 and describing congenital hindbrain disorders, consists of downward herniation of the cerebellar tonsils through the foramen magnum at the level of the cranial base. Patients commonly present with a symptom complex involving some combination of posterior cervical pain, headache, ocular or neurootological disturbances, posterior fossa abnormalities, and spinal cord dysfunction. Diagnosis is rather easily verified on routine MR imaging studies, and treatment is rendered through suboccipital decompression involving a C-1 laminectomy, thereby liberating the displaced tonsils from their stenotic bone confines.5,6,14

Among the many complications associated with decompression of a Chiari I malformation, cervical instability has received little attention in the literature. In an account of their experience managing 40 cases of Chiari II malformation, 20 of which were treated using suboccipital decompression, Aronson and colleagues demonstrated a significant increase in translation between C-2 and C-3 as well as angulation between C-3 and C-4 in the surgically treated group. Moreover, the authors also found 19 of the 20 surgically treated patients to possess some degree of cervical instability postoperatively. In contrast, no patient in the non–surgically treated group was found to have cervical instability, data confirming the lack of effect the malformation itself has on spinal stability. In an early paper describing kyphotic deformity arising in children who had undergone multilevel laminectomies, Cattell and Clark hypothesized that cervical instability in this setting can be attributed to muscular imbalance, abnormal growth patterns, and ligamentous stretching. In an even more compelling report, Gangemi and coworkers specifically studied the effect of posterior fossa surgery and upper cervical laminectomy on cervical stability in children. All patients had undergone suboccipital craniectomy, and the independent variable of interest was the inferior extent of cervical laminectomy. Of the 98 cases reported, cervical instability occurred in 3% of the patients who had undergone laminectomy of C-1, 22% of those who had laminectomies involving C-1 and C-2, and 100% of those who had undergone laminectomies of C-1 through C-3.

In addition to the generation of cervical instability following suboccipital decompression of a Chiari I malformation, one must consider the mechanism of atlas fracture in this context. Clearly, the two are intimately related from a biomechanical perspective. In his initial manuscripts elucidating the details of fractures involving the first cervical vertebra, Sir Geoffrey Jefferson attributed the bone disruption to an axial load force transmitted from the occiput to the lateral masses of C-1. Structures that are paramount in absorbing an axial load and resisting radial displacement of the lateral masses include the anterior arch, posterior arch, and transverse atlantal ligament. Following suboccipital craniectomy and C-1 laminecto-

Fig. 2. Left: Preoperative sagittal MR image of the craniocervical junction demonstrating the tonsillar herniation through the foramen magnum. Upper Right: Preoperative axial CT scan demonstrating the anteriorly displaced C-1 fracture. Lower Right: Postoperative axial CT scan revealing the healing anterior fracture.
my, a patient’s ability to withstand an axial load is undoubtedly diminished because the posterior arch is no longer intact. Moreover, the muscular, ligamentous, and fascial layers through which the surgeon must traverse during an upper cervical laminectomy no longer confer the same degree of stability to the atlas after having been breached.

Unlike classic atlas fractures in which the mechanism of injury is axial loading, a C-1 fracture following subcoccipital decompression cannot be solely attributed to this (note that neither of the patients reported in the current study had sustained a significant axial load). Nonetheless, we hypothesize that axial loading does indeed play a role in the pathological origins. The rarity of an atlas fracture in this setting might reflect not only its uncommon occurrence, but also its rare recognition because of spontaneous healing or an absence of symptoms in most cases.

Patients with atlas fractures can usually be treated quite effectively by using external orthotic immobilization in a hard cervical collar provided that the injury is stable. Open surgical fixation of C-1 fractures, by contrast, is usually reserved for patients in whom overt spinal instability is present based on traditional radiographic parameters or when failure of nonsurgical therapy persists. Criteria for evaluating the likelihood of transverse ligament disruption proposed by Spence and colleagues is easily applicable during the initial injury assessment; however, it has become our practice to obtain an MR image in most cases to assess transverse atlantal ligament integrity more directly prior to determining the optimal treatment strategy. Because atlas fractures commonly occur in conjunction with other cervical injuries, namely axis fractures, the indications for surgical intervention can also depend on the location and type of concomitant injuries. It has been our practice to perform open surgical fusion in cases in which a 3-month trial of external immobilization fails to relieve posterior cervical pain or eliminate cervical spinal instability, should either of the two be present. In addition, we perform surgery in patients in whom cervical instability subsequently develops, despite external immobilization.

Conclusions

Fracture of the atlas, which commonly results from an axial loading force, can occur in the postoperative period following C-1 laminectomy after decompression of Chiari I malformation. The mechanism underlying atlas fracture in this setting appears to be related to, but not entirely the result of, axial load forces, given that neither of the patients presented here had a history of such a mechanism. Of the several causes of postoperative neck pain in patients who have undergone decompression of a Chiari I malformation, an atlas fracture is important to consider in the differential diagnosis. Treatment of atlas fractures, which usually involves external immobilization with a hard cervical collar or a halo orthotic, is aimed at recovering bone continuity and stability in the upper cervical spine and craniovertebral junction. In patients who fail to heal despite the use of an external orthosis or in those who display evidence of spinal instability at the outset, a procedure aimed at achieving surgical fusion should be strongly considered, with or without the use of adjunctive spinal instrumentation.

References