Craniovertebral anomalies: Role for craniovertebral realignment

Atul Goel
Department of Neurosurgery, Seth G. S. Medical College and K. E. M. Hospital, and Lilavati Hospital and Research Centre, Mumbai, India.

Craniovertebral anomalies present an array of complex bony malformations leading to a range of symptoms secondary to neural compression, mal-alignment of bone and subtle or manifest instability. The treatment of these malformations is a challenging problem. Whilst reduction and fixation in the reduced position of mobile and reducible atlantoaxial dislocation is the standard and accepted form of treatment, the treatment of anomalies like basilar invagination, syringomyelia and ‘fixed’ atlantoaxial dislocation remain under discussion. Restoration of the distorted alignments without any bony decompression, dural or neural manipulation is possible and probably an ideal form of treatment in a select group of these anomalies.

On the basis of the literature review and our own study, we have observed that the craniovertebral maldevelopment in basilar invagination is due to a reduced length of the clivus (i.e., the sphenoid part of the clivus is formed relatively normally, whereas the occipital part is formed incompletely) and platybasia, occipital condylar hypoplasia, non-formation or inadequate formation of the occipitoaxial joint, and frequently, occipitalization of the atlas. Fusion of the atlantoaxial joint, and C2-3 spinal elements and a range of Klippel-Feil spinal abnormalities are also frequently associated. The entire complex of the odontoid process, the atlas, and clivus is rostrally located, and effectively the volume of the posterior cranial fossa is reduced. Partial or complete assimilation of the atlas is an important and frequent component of the mesodermal maldevelopment. Basal mesodermal maldevelopment will result in rostral positioning of the plane of the foramen magnum and significantly severe basilar invagination if measurements are taken on the basis of parameters laid down by Chamberlain. However, the tip of the odontoid process will remain below both Wackenheim’s clival line and McRae’s line of the foramen magnum in a large percentage of cases.

Chiari malformation and related pathological events could be primarily attributable to maldevelopment of the occipital bone and over crowding of the normally developed cerebellum within a smaller posterior cranial fossa. There is usually no demonstrable structural abnormality of the brainstem, cerebellar hemisphere or fourth ventricle suggesting that the neural development in these patients was unaffected in the embryonic dysgenesis. Chiari malformation or tonsillar herniation appears to be the result of the presence of normal cerebellar mass in a smaller posterior cranial fossa. Long-standing pulsatile pressure of the herniating tonsil to the brainstem as a cause of formation of syringomyelia cavitation has been discussed. Essentially, it appears that syringomyelia is a tertiary event to the primary basilar invagination and secondary Chiari malformation. We had suggested that the treatment strategy in such cases should be directed towards increasing the posterior cranial fossa volume by foramen magnum decompression, which is the cause of Chiari malformation and subsequent syringomyelia. Opening of the posterior cranial fossa dura, resection of the cerebellar tonsils, arachnoidal sectioning and syringostomy can be avoided. The theory that dura or dural bands can act as a compressive factor appears unacceptable and does not correlate with the pathogenic events. Dura is an expansile structure and is unlikely to be a compressive factor. Our observation is that if the syringomyelia is treated without dealing with the pathogenetic factor, more often than not the outcome will be poor.

Treatment of selected cases of congenital basilar invagination: We had discussed the subject of basilar invagination and had presented a classification for these anomalies based on the presence or absence of the Chiari I malformation. On the basis of the possible pathogenetic factors, we had suggested a specific treatment protocol for each of the two groups. With our improved understanding of the subject, we re-classified basilar invagination into two groups, Group A and Group B, based on parameters that determined an alternative treatment strategy. In Group A basilar invagination there was a ‘fixed’ atlantoaxial dislocation and the tip of the odontoid proc-
ess 'invaginated' into the foramen magnum and was above the Chamberlain line, \textsuperscript{7} McRae line of foramen magnum \textsuperscript{2} and Wackenheim’s clival line.\textsuperscript{4} The definition of basilar invagination of prolapse of the cervical spine into the base of the skull, as suggested by von Torklus,\textsuperscript{9} was suitable for this group of patients. We reviewed our entire series of cases with basilar invagination and identified that Group A basilar invagination constitutes approximately 60\% of all cases of basilar invagination.

The majority of patients with Group A basilar invagination had a history of minor to major head injury prior to the onset of the symptoms. The pyramidal symptoms and affection of the kinesthetic sensations formed a dominant component. Spinothalamic dysfunction was less frequent. Neck pain as a major presenting symptom was in approximately 75\% cases. Torticollis was present in approximately 50\% cases. The analysis of the clinical features suggests that the symptoms and signs in this group of patients were a result of brainstem compression by the odontoid process. The radiological features suggested that the odontoid process resulted in direct compression of the brainstem. Analysis on the basis of Chamberlain’s line showed that the basilar invagination was mild to severe in these cases. Modified omega angle measurements suggested that the odontoid process had tilted towards the horizontal rather than rostrally.\textsuperscript{10} The patients had a 'fixed' atlantoaxial dislocation and no atlantoaxial mobility could be identified on dynamic radiology in any case.

The standard and most accepted form of treatment of Group A basilar invagination is a transoral decompression. The majority of authors recommend a posterior occipitocervical fixation following the anterior decompression. It appears to us that the atlantoaxial joint in such cases is in an abnormal position as a result of congenital abnormality of the bones and progressive worsening of the dislocation is probably secondary to increasing 'slippage' of the atlas over the axis. The slip of the atlas over the axis appears to be accentuated by the event of trauma. With our experience in handling the atlantoaxial joints, we have realized that the joint in these cases is not 'fixed' or 'fused' but is mobile and in some cases is hypermobile, and is probably the prime cause for the basilar invagination. The history of trauma preceding the clinical events, predominant complaint of pain in the neck and improvement in neurological symptoms following institution of cervical traction suggests 'vertical' instability of the craniovertebral region.

We had earlier attempted to reduce basilar invagination by performing occipitocervical fixation following institution of cervical traction.\textsuperscript{1} However, all the four cases treated in this manner subsequently needed transoral decompression as the reduction of the basilar invagination and of the atlantoaxial dislocation could not be sustained by the implant. Wide removal of the atlantoaxial joint capsule and articular cartilage by drilling and subsequent distraction of the joint by manual manipulation provided a unique opportunity to obtain reduction of the basilar invagination and of the atlantoaxial dislocation. The joint were maintained in a distracted and reduced position with the help of bone graft and spacers. The subsequent fixation of the joint with the help of interarticular screws and a metal plate provided a biomechanically firm fixation.\textsuperscript{11-13} The fixation was seen to be strong enough to sustain the vertical, transverse and rotatory strains of the most mobile region of the spine. Following surgery, the alignment of the odontoid process and the clivus and the entire craniovertebral junction improved towards normalcy. The tip of the odontoid process receded in relationship to Wackenheim’s clival line, Chamberlain’s line and McRae’s line suggesting reduction in the basilar invagination. The posterior tilt of the odontoid process, as evaluated by modified omega angle, was reduced after the surgery. We could obtain varying degrees of reduction of the basilar invagination and atlantoaxial dislocation. The extent of distraction of the joint and the subsequent reduction in the basilar invagination was more significant in younger than in older patients. The procedure is technically demanding and anatomically precise,\textsuperscript{14-16} but if it is learned adequately and performed successfully, the neurological outcome is extremely gratifying. Posterior fossa bony decompression appears to be an ideal form of treatment for Group B basilar invagination.

The procedure of distraction of the facets of the atlas and axis, placement of spacer and bone graft within the joint and its subsequent fixation was also used to treat cases of basilar invagination related to rheumatoid disease\textsuperscript{17} and cases with fixed atlantoaxial dislocation without any basilar invagination.\textsuperscript{18}

**Treatment of selected cases of syringomyelia:** We had classified syringomyelia in three groups and had suggested a specific treatment protocol on the basis of the possible pathogenetic factors.\textsuperscript{17} The complex of basilar invagination, Chiari 1 malformation and syringomyelia is relatively common and there are multiple reports on the subject. Association of fixed atlantoaxial dislocation with the latter group is less common but not rare. Such cases are generally treated by either anterior transoral or posterior foramen magnum bony decompression. The indications and the need for opening of the dura and manipulation of the arachnoid membrane, tonsils and obex and draining of the syrinx cavity are currently under debate. There are no reports in the literature identifying the need for a specific fixation procedure for cases of fixed atlantoaxial dislocation where it is associated with basilar invagination in the presence of syringomyelia.\textsuperscript{19}

It was observed that cases of syringomyelia where there was ‘fixed’ atlantoaxial dislocation with or without the association of basilar invagination and Chiari malformation constituted a discrete pathologic group. It was observed that in this group, the patients were relatively young, neck pain formed a part of the symptom complex and the motor symptoms and
ataxia were more prominent symptom. We observed that an attempt could be made to realign the bones in the craniovertebral junction in these cases, without resorting to any bony or dural decompression or neural manipulation of any kind. The fact that there was a remarkable clinical improvement following the reduction of the atlantoaxial dislocation and of basilar invagination, it appears that the complex of atlantoaxial dislocation, basilar invagination and syringomyelia are probably secondary to the primary craniovertebral instability.

References