

Basilar Invagination, Chiari Syndrome and Critical Compression of the Bulbomedullary Junction: Genesis and Pathophysiology of an Entity not Clearly Understood

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Abstract

Introduction: Basilar invagination is a complex malformation of the cranio-cervical junction where treatment consensus has not been reached and the therapeutic approach must be individualized according to the case and the associated anatomical variants. In the most serious cases, an adequate therapeutic approach can determine significant or total regression of neurological symptoms.

Objective: The objective of this review is to arrive at the analysis of the treatment algorithms based on an adequate understanding of the anatomy, anatomical variants and conceptualization as close as possible to the most correct diagnosis for the application of the best treatment method.

Methods: A bibliographic review of the main existing works on malformations of the craniocervical junction was carried out, placing emphasis on anatomical morphological, biomechanical and physiological and radiological diagnosis.

Conclusion: The adequate conceptualization and classification of the pathology allows the correct interpretation of the imaging findings and clinical findings as a reference framework for treatment. However, each patient has individual characteristics and the concept itself constitutes a continuum of malformations that must be resolved by different surgical routes, at different times if necessary and supported by different support technologies.

Keywords: Basilar Intussusception; Cranio-Cervical Bone Malformation

Introduction

Odontoid recession is a malformation of the hinge sometimes described in association with basilar impression or platybasia. However, recognizing the exact morphology and that this entity may present in isolation and cause significant compression of the brainstem and cervical spinal cord helps to define a better treatment strategy [1]. The congenital alteration can occur with several combinations that can be derived from alterations in nutrients and vitamins during pregnancy as a probable cause. This to be developed during the first phases of the formation of the embryo [1]. The pathogenesis of Chiari malformation is not clearly recognized or poorly understood. In many cases, it is associated with bone malformations and basilar intussusception. In cases where there is no clear cause or it is associated with bone malformations, the Chiari malformation is generally considered to be part of a pathological complex or a congenital malformation. Basilar intussusception is a common bone disorder and syringomyelia is a malformation of associated soft tissues. Syringomyelia is considered a mistake in embryogenesis. Syringomyelia is frequently associated with a blockage of the flow of the CSF pathways at the level of the foramen magnum and herniation of the tonsils. A clear morphological differentiation between platybasia, basilar impression, basilar invagination, and odontoid recession is important, although all these alterations generally lead to deformity and compression of the brainstem. Platybasia classically defined as the flattening of the base of the skull with an alteration of the basal angle of Boogard that normally oscillates between 115 and 140°, when it exceeds the latter it is spoken of platybasia, but it is important to know that it can be accompanied by a basilar impression, which can determine the appearance of important symptoms. IMB is an invagination of the bony

contour of the foramen magnum into the posterior fossa with a reduction in its capacity, giving the skull base a dome shape opposite to the normal one (convexobasia). This abnormal arrangement is still attributed to an early synostosis of the sphenoccipital suture [2,3]. The association of these entities with Down Syndrome, Chiari Malformation, Syringomyelia and Klippel Feil Syndrome is high and may determine different treatment strategies. Other associated acquired pathologies are rheumatoid arthritis.

Symptoms of compression of the craniocervical junction can be nonspecific and difficult to locate. The diagnosis can often be delayed and be confused with demyelinating diseases or other types of pathologies. Ischemic symptoms can be triggered if there is involvement of the basilar or vertebral artery, PICA, or anterior spinal artery [4]. Due to the drastic nature of the condition and the potentially disabling or even fatal, the treatment is usually surgical, and various approach strategies may be presented. Here we present a case of a patient with an odontoid recession documented with X-ray, CT and MRI with its corresponding clinical correlation. This review is not intended to focus on the various types of treatment proposed, however it does intend to clarify the possibly common origin of several Syndromes or entities considered isolated and that could potentially be treated as a whole depending on the pathological associations and clinical findings.

Embryology

The aspects and chronology of clear embryological development play a role in relation to this pathology or this continuum of malformations that form this compressive neural syndrome. At day 15 of embryo development, the primitive streak that forms the primitive nodule can be identified. The notochord forms a central cluster of cells that separates the ectoderm from the endoderm and on the sides the mesoderm is observed that unfolds and separates the endoderm from the ectoderm, except for 2 points: the oral membrane in the cephalic portion and the cloacal membrane at the end of the notochord. The notochord fuses with the endoderm on day 21 of development, determining a wide communication between the amniotic sac and the Yolk sac, forming the neurenteric canal. On day 21 this canal is completely obliterated with the development of the notochord. Between days 17 and 19 in the process of neurulation, the mesoderm forms two columns on the sides of the midline, the paraaxial mesoderm. This mesoderm gives rise to the somites that begin to segment on the 20th day in the occipital region and on the 35th day they form 42 somites. The dorsolateral components of the mesoderm will form the dermatomes, the medial cells will form the dorsal musculature, and the ventromedial cells will form the sclerotomes. It is these latter cells that with their migratory capacity that move around the notochord and the neural tube to form the vertebrae and discs. The development of the spine cannot be analyzed separately in relation to the development of the neural tube at day 23, especially due to the migratory situation of the mesenchymal cells derived from paraaxial mesoderm. The formation of the vertebrae and intervertebral discs is very complex but explains important findings in the adult anatomy. The segmentation of sclerotome derived from a somite evolves between an area of less cellularity in the upper part and an area of greater cellularity in the lower part of the segment with a marked condensation of cells in the upper part that is destined to form the disc and the fibrous ring. The less dense component subsequently fuses with the less dense component of the sclerotome forming the center of the vertebra. During this process the notochord that has become the vertebral body and persists in the vertebra formed as the mucoid stria. In the region of the disc it persists and many of the notochord cells migrate to this area to become the nucleus pulposus and the disc.

Brain development is complex and requires mentioning it only because of the association with some pathologies of the cranio-vertebral union. On day 24, the neural tube begins to close and the 3 deprived vesicles rapidly form. When the embryo begins to flex, the first flexion begins in the cervical spine and brainstem. The first flexion begins between the bridge and the midbrain and the rapid development of the cerebral hemispheres gives rise to the brainstem. From a practical point of view, the most important flexion is the third, at the pontine level, which flattens the neural tube at this point and begins the formation of the fourth ventricle. This occurs between the 30th and 36th day [2].

History

The characteristic definition of an Arnold Chiari Malformation consists of various degrees of ectopy of the cerebellum and medulla through the foramen magnum. A lesser degree may consist of the tonsils resting in the cervical canal at the C1 level. In a more serious case, part of the bulb and elongated tonsils extending to the middle of the cervical canal may be the characteristic. Historically there was a common association between the more severe forms and spina bifida and it has been suggested that the cerebellum is pushed down by hydrocephalus or is pushed down by the developing malformation which prevents the spinal cord from herniating cephalad. The other degrees of Arnold Chiari are important to the extent that there is syringomyelia associated with it or not. It is not yet clear what is the dynamic relationship or the coincidence between the different malformations [3]. The multilocular nature of some cavities is inconsistent with a simple balloon effect in many cases, making the decompression of the prolapsed structures not always successful in the treatment of syringomyelia, although in many cases the progression may be delayed. Bony abnormalities in the region of the foramen magnum and cervical spine occur by an uncertain embryogenic mechanism. This can lead to a small posterior fossa, with basilar invagination and the odontoid invading the foramen magnum, or a vertebral fusion with Klippel Feil syndrome. All these abnormalities can occur with a short neck. These abnormalities can produce complications at the posterior fossa level in later stages in middle ages. For example, Klippel Feil Syndrome can determine degenerative changes in the remaining segments of the neck leading to irritation of nerve roots in the spinal cord. In 2004 Goel, *et al.* [23] proposed in a group of cases with basilar intussusception (Group A), a manual distraction of the atlas and axis facets, resulting in a realignment of the craniovertebral junction [3,23]. They identified in group A a basilar invagination, the atlantoaxial joint was unstable but not fixed or irreducible. At that time, this group advocated the use of intra-articular spacers, adding bone graft to distract and stabilize the facets and realign the odontoid process inferiorly in this final position. Currently the same group proposes that even more important than realignment is firm stabilization and fusion of the bone in the atlantoaxial joint. In group A, of patients with basilar intussusception, transoral decompression and resection of the odontoid process were previously recommended. Treatment has changed to decompression and stabilization in group A cases. This concept has relegated transoral surgery to an almost historical anecdote [29]. In cases with Group Basilar Invagination where fixed atlantoaxoid instability is considered, the recommended treatment is decompression of the foramen magnum [27].

There are treatment algorithms in relation to the old concepts that cannot be completely discarded, especially for malformations of the hinge, and it has been established that if a malformation is reduced, a posterior fixation should be chosen. If the malformation is not reduced, it should be considered whether the compression is anterior or posterior, on which the surgical approach will depend [6]. Traction reduction is primarily tested in patients with rheumatoid arthritis, infections, neoplasms, and bone softening diseases [4,6]. Summarizing the treatment approach, the treatment algorithms for flap malformations should be used according to the classification of the abnormality. At this point, Dickman, Ronderos and Sonntag [13,19] established that if a malformation is reduced, a posterior fixation should be chosen. Traction reduction has been practiced primarily in patients with rheumatoid arthritis, infections, neoplasms, and bone softening diseases [7-12]. If the malformation is not reduced, it should be considered whether the compression is anterior or posterior, on which the surgical approach will depend. In cases of ventral compression and the presence of compression by inflammatory or fibrous tissue, a posterior decompression must first be performed including C1-C2 and occipital cervical fixation, due to the possibility of critical instability between the first and second intervention [6]. If there is no improvement or if compression persists in the control image, ideally with MRI, a transoral odontoidectomy should be performed [6]. On the other hand Goel [8] proposes an algorithm depending on 2 groups. Group 1 or "unstable" where the effective reduction or not, must be subject to fixation by posterior means, whether it is mobile or not. In group 2 considered stable, a subsequent decompression is sufficient. Regarding the posterior approach, there is no consensus regarding the use of dural plasty, it was initially described by Goel in 1985. Subsequently, and with the increase in cases and controls, the same group recommends treating the 2 variants, both type A and type B, as alteration of C1-C2 stability, and changes the focus from decompressive surgery to stabilizing surgery [41].

Discussion

Chiari malformation is rarely associated with posterior fossa tumors, hydrocephalus, and hydrosyringomyelia [9,10]. The symptoms can be multiple and the diagnosis can be deferred due to the nonspecific nature and the difficulty in locating the main pathology. These findings are often confused with demyelinating diseases. Ischemic symptoms may occur if the vertebral basilar system, the PICA arteries, or the anterior spinal artery are involved. Alteration of the cranial nerves and instability may also occur, resulting in neck pain, occipital neuralgia, radicular pain, or worsening of neurological symptoms [11]. To this can be added defense mechanisms such as the contracture of the muscles of the neck that often even determine torticollis [15,16].

Congenital malformations of the cranio-cervical junction are generally not static and can be reduced with traction [17-19]. According to the groups studied by Goel, *et al.* The main fact resides in an instability of C1-C2, which is the joint where the main movements of the cranio-cervical union occur [20,21]. In this sense, the casuistry collected and analyzed by Goel, *et al.* For 30 years, determining that the C1-C2 joint is the center of movement and also the center of stability of the craniocervical junction [22-25]. The atlanto axoid dislocation is the main fact, little understood until recently and considered a fixed abnormality, which can now even be considered unstable even though there is a normal distance between the atlas and the odontoid [22]. Chiari malformation and herniation of the cerebellar tonsils in the foramen magnum appear to be the protective mechanism in atlantoaxial instability [23]. This could be a natural "air bag" that prevents compression of the trunk on the bony structures. Syringomyelia would develop to reduce pressure on the intracranial and spinal structures [1]. Basilar invagination has long been associated with the presence of a fixed or irreducible atlantoaxial dislocation. Because the malformation was considered fixed, bone decompression to provide additional space for neural structures was the treatment of choice [1,24].

There is a new classification proposed by Goel, it would be 3 groups [25,26]. Type A facet dislocation when the facet of the atlas is dislocated and situated anterior to the facet of the axis. This determines a postero-superior displacement of the odontoid and is associated with atlanto-axoid dislocation and basilar invagination. There is an increase in the atlanto-axoid space and the odontoid displaces posteriorly in the spinal canal. The result is acute or subacute compression of neural structures. In type B, the facet of the atlas dislocated behind the facet of the axis, in this form of instability the odontoid did not move posteriorly in the canal and the distance to C1-C2 was not significantly altered [3]. Type C with facet dislocation occurs when there is no misalignment of the facets and instability is identified only on the basis of clinical and intraoperative observations. Instability in some cases can be central or axial [26-28].

Chiari malformation and herniation of the cerebellar tonsils in the foramen magnum appear to be the protective mechanism in case of atlantoaxial instability [29]. Basilar invagination has long been associated with the presence of a fixed or irreducible atlantoaxial dislocation. Because the malformation was considered fixed, bone decompression to provide additional space for neural structures was the treatment of choice [30]. Patients with tumors of the craniocervical junction or infections may require transoral surgery for resection surgery or debridement of their pathology in addition to neural decompression. If the instability is associated with the pathology or it develops postoperatively, it is indicated to do a second stage posteriorly to fix the craniocervical segment. Transoral anterior odontoidectomy provides a direct anterior approach that avoids manipulation and maneuvering over a critically affected area by compression on the medulla and medulla, the cranial nerves, and important vascular structures [31-35]. Compared to the superior retropharyngeal approach for the craniocervical junction, the advantage is that in the latter the cranial nerves are manipulated, branch vessels of the external carotid artery are sacrificed, and the pharynx is extensively mobilized. These maneuvers result in significant dysphagia that can be avoided with the transoral approach. The retropharyngeal approach has the advantage that, being extramucosal, it can allow the placement of a bone graft or titanium implants with a very low infection rate [36-40].

Basilar intussusception is a complex of alterations, which can be considered a continuum and even a mixture of possibly congenital morphological alterations, associated with better defined atlanto-axial, axial or central instability. Musculoskeletal alterations in basilar

invagination present with short neck, short spine, torticollis, platybasia, bone fusion, such as C2-C3 fusion, Klippel-Feil, bifid posterior arch of the wings, os-odontoidum and manifestations neurological symptoms in the form of Chiari, external syringomyelia, syringohydromyelia, external syringobulbia and syringobulbia [42].

Conclusion

The stages of embryonic development from day 17 to 29 with the formation of the neural tube and the concomitant development of the lateral mesoderm and the formation of the axial skeleton, especially the vertebral column, make the development of congenital malformations such as Chiari Malformation associated with disorders perfectly possible. musculoskeletal. The excessive presence of fluid inside and outside the nervous system is known to constitute a defense mechanism of the neural tissue against external noxas in such a way that a trauma to the odontoid against the brainstem or medulla is minimized in terms of its impact [42]. The therapeutic approach consolidated for years deserves at least a review of the pathophysiological mechanism in light of the radiological findings that include a good study of the planar craniocervical junction CT, functional resonance or the correct interpretation of functional Ap x-ray images. and lateral. In addition, giving the maximum value to surgical findings and what seems to be a tremendous turnaround in the approach to surgical treatment and its role in the resolution of this malformative complex.

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