The first description of Chiari I malformation with intuitive correlation between tonsillar ectopia and syringomyelia

Historical vignette

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Few are familiar with the neurological contributions of the German pathologist Theodor Langhans. Even fewer are aware of his significant and early contributions to the study of what is now known as the Chiari I malformation. In at least 4 cases, Langhans described the association between tonsillar ectopia and syringomyelia. Moreover, this early pioneer speculated that there was a cause and effect with hindbrain herniation resulting in improper flow at the craniovertebral junction and consequent development of syringomyelia. These cases were reported prior to Hans Chiari’s descriptions, and Langhans’ theory of impeded foramen magnum flow as a cause of syringomyelia was novel and preceded the current understanding of this mechanism by almost a century. The authors discuss the life of Langhans and translate excerpts from his 1881 work regarding tonsillar ectopia and syringomyelia. (DOI: 10.3171/2010.12.PEDS10579)

Key Words • pathology • hindbrain • Chiari malformation • syringomyelia

The 4 types of Chiari malformations represent varying degrees of involvement of rhombencephalic derivatives. Three of these (Types I–III) have herniation of these structures outside of the posterior fossa. Currently, there is no consensus on the exact pathogenesis or treatment for any of these malformations. Although the Chiari classification is helpful in categorizing most cases, this scheme most likely does not represent a precise continuum of the same disease process and does not adequately define all forms of hindbrain herniation encountered.

Chiari’s Contributions

In the early 1890s, the Austrian, Dr. Hans Chiari, professor of pathological anatomy at the German University in Prague, used autopsy specimens to describe 4 congenital anomalies later termed the Chiari malformations (Types I–IV). Since this initial description, our understanding of these lesions has evolved into several main concepts: 1) the Chiari malformations are separate anatomical entities, all of which involve the hindbrain; 3) of the 4 consist of a type of hindbrain herniation, whereas the fourth consists of cerebellar hypoplasia or aplasia; 2) the herniation occurs as an imprecise continuum ranging from mild tonsillar herniation through the foramen magnum to severe cerebellar and brainstem herniation into a posterior encephalocele leading to syringomyelia, medul-lary compression, and ischemia; 3) despite their common anatomical location, the 4 anomalies have distinct clinical-atomal features; and 4) although the large majority of cases are congenital, cases of acquired Chiari I malformation have been reported.

Other Early Reports of Hindbrain Herniation

Chiari was not the first to observe and report on Type II malformations. In Observationes Medicæ, the famous Dutch physician and anatomist Nicholas Tulp (1593–1674) described an individual with myelodysplasia and may have referred to hindbrain herniation (Chiari II malformation), although this is controversial. In 1829, Cruveilhier (1791–1874) of Paris described a case of myelomeningocele in which “…the considerably dilated cervical region contained both the medulla oblongata...
and the corresponding part of the cerebellum, which was elongated and covered the 4th ventricle, itself enlarged and elongated.6 Again, this description by Cruveilhier was of the entity that would become known as the Chiari II malformation. Decades later, in 1883, John Cleland (1835–1925) of Scotland reported a single case of myelodysplasia in which there was hindbrain herniation and hydrocephalus.3 In 1894, Julius Arnold (1835–1915), professor of anatomy at Heidelberg, described a single myelodysplastic case with hindbrain herniation and no hydrocephalus.3 Little attention was given to the posterior fossa abnormalities in this report, but 2 of his students seized this opportunity to immortalize their professor by affixing the moniker “Arnold-Chiari malformation” to this condition.3 In the end, however, it was the significant contributions of Chiari that shed the most light on these forms of hindbrain herniation; thus, referring to them as Chiari malformations is appropriate.

### Langhans and his Descriptions of Chiari Malformation and Syringomyelia

Theodor Langhans was born September 28, 1839, in Usingen (Nassau), Germany, and studied under Henle in Göttingen and von Recklinghausen in Berlin (Fig. 1). He attended medical school in Heidelberg and, in 1864, completed his medical degree thesis on the structure of tendons in Würzburg. He was also a student under such names as Virchow, Trauber, and Frerichs.1 He served as assistant to von Recklinghausen until 1867. In Marburg, he collaborated with Lieberkölin and Wagner on anatomical research. He was later made Professor Ordinarius in Giessen and then moved to Switzerland in 1872 where he was appointed Professor and Chair of Pathological Anatomy in Bern, succeeding Klebs. He described the giant “Langhans cells” of tuberculosis lesions in 1867 and his name was given to these cells by the famous Carl Weigert in 1881.2,5 Langhans with the physician Sahli and the surgeon Kocher formed a triumvirate, which made the medical school at Bern famous.1 During this time, Langhans also described the formation of pigments and the histology of the breast, placenta, cornea, and testicle.2 He went on to study goiter and cretinism, and the distribution of glycogen in normal and diseased organs, and is also eponymously remembered for his descriptions of the chorionic epithelium. In his 1881 publication Über Höhlenbildung im Rückenmark als Folge Blutstauung (translated as “Regarding cavity creation in the spinal cord following a change in the cerebellar cavity, I could not find a cause for the increase in pressure; but great pressure on the pons and medulla oblongata from above was indeed apparent. Upon dissection of the cerebellum, nothing was of note except for an obvious/significant development of both tonsils, which protruded down in the form of two symmetrical pyramidal tumors and pushed the medulla oblongata in a frontal direction at almost a right angle.

The formation of the cavities, according to my observations, was connected to other changes in the central nervous system, more specifically to changes in the cerebellar cavity, which must have impeded the circulation to a great extent. The increase in pressure in the cerebellar cavity will hinder or greatly impede the outflow of blood and cerebral spinal fluid.

In all cases, the ventral part of the spinal cord is affected and if at all, only a small portion of the dorsal part. The cavities do not start in the medulla oblongata at the calamus scriptorius or in the upper 1-2 cm of the spinal cord.

The direction in which the central canal extends is constant – to the side and posteriorly. In my opinion, the decisive factor for this is the consistency of the white matter. The cavity creation starts there where the increased pressure, which exists in the cerebellar cavity stops and, therefore, a diverticulum can only occur toward the area of less pressure.
According to my theory, a diverticulum is more likely to occur than a widening of the central canal, because the development of the diverticulum in the dorsal part meets less resistance than a central expansion.

These descriptions are striking for several reasons, including Langhans' first describing pathological tonsillar ectopia and hypothesizing that this obstruction at the foramen magnum results in development of syringomyelia. Additionally, he clearly recognized the fact that syringomyelia normally does not include the first segment of the cervical cord. Lastly, Langhans realized that fluid accumulation within the spinal cord could occur via dilatation of the central canal or outside of this region (Fig. 3).

Name Confusion

Interestingly, Pritchard et al. have reported that the similarities in surnames between Theodor Langhans and his contemporary Paul Langerhans (1847–1888) have resulted in clinical errors in diagnosis with consequences. Langerhans is remembered for the islets that he discovered in the pancreas and the antigen-presenting cells of the dermoepidermal junction. Both men published in Virchow's Archives in 1868 and both died of tuberculosis. Two years prior to his death on October 22, 1915, Langhans retired as chairman of Pathological Anatomy in Bern.

Although the association of Chiari I malformation and syringomyelia would not be commonly used until the late 20th century, it was Langhans in the 19th century who proposed this cause and effect. Therefore, appropriate recognition for this association should be given to this early pioneer who, with Chiari, helped provide us with details of hindbrain herniation that are still in use today.

Disclosure

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

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