From the Pages of History

About Chiari, Arnold & The Malformations

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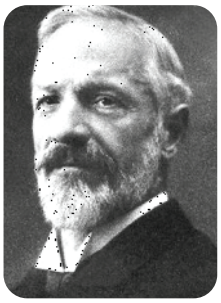

Hans Chiari (1851-1916)

The congenital hindbrain herniations are known as Chiari malformations and include 4 types. The type II Chiari malformation is better known as Arnold-Chiari malformation. Here is a brief historical account of Chiari and Arnold and how these malformations came to be described.

Hans Chiari (1851-1916) was born in Vienna. He was the son of Johan Baptist Chiari, who was a physician and is credited with the description of prolactinomas. Hans Chiari was closely associated with the then renowned Pathologist, Karl Rokitansky during his medical education at Vienna. After completing medical education, Chiari was associated with Richard Ladislaus Heschlat the Institute of Pathology at Vienna, who succeeded Rokitansky. In 1882 Chiari joined the German University at Prague, where most of his accomplishments came from. His major contributions have been: the first description of features of choriocarcinoma, description of the thrombosis of hepatic vein (Budd-Chiari syndrome) along with the British physician, George Budd, the role of the carotid artery plaques in the pathogenesis of thrombosis, the symptoms of fistula between aorta and esophagus caused by ingestion of foreign body or gunshot injury, the first description of traumatic pneumocephalus and demonstration of the CSF fistula in the base of anterior cranial fossa and description of a new transnasal approach to the pituitary gland lesions. He also demonstrated communication between syrinx and the central canal of spinal cord. Chiari first published his works regarding hindbrain malformationsin the journal Deutsche Medizinische Wochenscriff in 1891and later in 1896. These were described in his paper titled "Ueber Veränderungen des Kleinhirns infolge von Hydrocephalie des Grosshirns"(which means "concerningchanges in the cerebellum due to hydrocephalus of the cerebrum") and these are presently called Chiari malformations[1]. The Chiari's type I malformation described by him first was a 17-year-old woman who died of typhoid fever and was found to have hydrocephalus,with "no symptoms referable to the cerebellum or medulla[1]."She had "peg-like elongation of tonsils and medial divisions of the inferior lobes of the cerebellum into cone shaped projections, which accompany the medulla oblongata into the spinal canal" without involving the medulla[1].

Julius Arnold (1835-1915) was a student of Rudolf Virchow and Nikolaus Friedreich at Heidelberg. In 1894, Arnold described a child with myelodysplasia without hydrocephalus, who had herniation of fourth ventricle and cerebellar tonsils and inferior vermis through the foramen magnum. Chiari's type II malformation, had similar description to Arnold's, was described as a "displacement of parts of the cerebellum and elongated fourth ventricle, which reach into the cervical canal"[1]. Subsequently Chiari redefined the type II malformations to include more hindbrain involvement, as a "displacement of part of the lower vermis, displacement of the pons and displacement of the medulla oblongata into the cervical canal and elongation of the fourth ventricle into the cervical canal[1]". In the year 1907, Schwalbe and Gredig, trained by Arnold in his laboratory, renamed Chiari type II malformation as Arnold-Chiari malformation.

Chiari type III malformation was of a severe variety, where there were "cervical spina bifida, partially absent tentorium cerebelli with prolapsed of the fourth ventricle and cerebellum into the cervical canal with hydromyelia communicating with the fourth ventricle[1]". In Chiari type IV malformation, there was no herniation of hind brain, but there was cerebellar hypoplasia associated with hydrocephalus. In 1906, Chiari moved out of Prague because of political tensions and joined University of Strasbourg, France. Chiari had published about 180 papers between 1876 and 1916. He passed away in 1916 due to a throat infection.

Thus, the congenital hindbrain herniations, which were first described by Hans Chiari, who grouped them into four types, should be called "Chiari Malformations". The term "Arnold-Chiari Malformation" should be applied only to the Chiari type II Malformationand not to the other types.

References

1) Tubbs RS, Oakes WJ. The Chiari Malformations: A Historical Context. In: Tubbs RS, Oakes WJ (eds.), The Chiari Malformations, DOI 10.1007/978-1-4614-6369-6_2, Springer Science+Business Media New York 2013: pp 5-11.