JBNC Brazilian Journal of Neurosurgery
Jornal Brasileiro de Neurocirurgia

Original

Brief historical note about Hans van Chiari (1851-1916)

Breve nota histórica sobre Hans Van Chiari (1851-1916)

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ABSTRACT

Hans van Chiari (1851-1916) was an illustrious physician who gave many contributions to the scientific knowledge of Medicine and medical history. This paper describes a brief historical note and his contribution along the history of Medicine.

Key words: *Chiari; Hans Van Chiari; Historical note; Bibliography.*

RESUMO

Hans van Chiari (1851-1916) foi um ilustre médico, que muito contribuiu para ciência do conhecimento médico e História da medicina. Esse trabalho descreve uma breve nota histórica e sua contribuição para a história da Medicina.

Palavras-chave: *Chiari; Hans Van Chiari; Nota histórica; Bibliografia.*

Brief History

In 1851, November 4th, Hans van Chiari was born in Vienna, and this Austrian physician contributed to the Medicine history. He was from a family of doctors, and his father, Johan Baptist Chiari (1817-1854) was a famous gynecologist in Vienna and Prague recognized for his paper on prolactinomas. His brother, Ottokar, was an Otolaryngologist [1].

Hans Chiari attended a Medicine course in Vienna and worked at the Vienna Pathology Institute as an assistant of a famous pathologist at that time, called Karl Rokitansky (1804-1878). In 1875, Rokitansky retired, and Richard Ladslau Heschl (1824-1881) assumed the management of the chair of Pathological Anatomy in Vienna; Chiari helped him until his death. Due to his performance, the attention to the details and cataloging pathologies at the Institute, Hans Chiari achieved great reputation. In 1877, he described for the first time some findings about choriocarcinoma. In 1882, he became an assistant pathologist physician at the German University, in Prague. One year after that he was nominated Superintendent of the Prague Pathological Museum and the majority of his work was developed while he lived in the city [1].

In a joint-work with the English physician George Budd (1808-1882), Chiari described the clinic and pathology of the hepatic vein thrombosis, that today is called

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Submitted Nov 7, 2017, Corrected Feb 22, 2018, Accepted Feb 25, 2018

as Budd-Chiari syndrome, a rare illness (affecting 1:100.000) [2]. This syndrome is characterized by portal hypertension and hepatomegaly, which follow the thrombosis in the hepatic vein, generating a liver drainage system obstruction[3]. In 1845, Budd described three cases of hepatic vein thrombosis; and, in 1899, Chiari performed the first pathological description of a liver with endoflebitis obliterans in hepatic veins[2]. The treatment for this syndrome is based on anticoagulation, regardless the prothrombotic disorder, also including percutaneous angiography, transjugular intrahepatic portosystemic shunt, and liver transplant[4]. About 25% of the patients remain asymptomatic after the treatment[5].

The pathologist Hans Chiari showed a special interest for the glands and wrote sufficiently about the autodigestive capacity of the pancreas. In the cardiovascular area, he described the connection between the Thebesian (coronary sinus valve) and Eustachian valves (inferior vena cava valve), besides the association of atherosclerotic plaques in the carotid artery and thrombosis. Chiari also described the association of symptoms with aortoesophagea[l] fistula due to foreign body ingestion or penetrating wound. Also, he gave an important contribution to the pituitary adenomas treatment, when he considered, in 1912, the transnasal surgical approach for these injuries[1].

However, it was in Neurology and Neuroanatomy that his papers became well known, when he first described brainstem and cerebellum malformations. In 1883, the probably first case reported on traumatic pneumocephalus description before the X-ray invention[1].

In 1891, he wrote an article called "On cerebellar changes from cerebral hydrocephalus" [2], describing a group of malformations caused by cerebellum changes, corresponding to the cerebellar tonsils elongation and the medial divisions of the cerebellum inferior lobe to the conic projections that accompany the spinal bulb in the spinal canal, called Chiari malformations. Julius Arnold complemented these findings in 1894, and in 1903, Schwalbe and Gredig [1,6] named the findings as Arnold-Chiari malformations in honor of those pathologists[7]. It is known that this pathology is one of the most important discoveries of Chiari; its prevalence is 0.4/1000 live births and corresponds to 3% of abortions[6]. Clinically, presentations can vary and, in 1965, Campbell[1,8] reported respiratory dysfunction associated with this malformation, characterized by nocturnal hypoventilation (the most common is sleep apnea) [9]. Moreover, the change in the circulation of the cerebrospinal fluid is another important aspect that this pathology can generate, which can be corrected after surgical treatment [10]. Imaging exams (Ultrasound or MRI) of the posterior fossa give the diagnosis[8], however, it can be often difficult, due to the varied neurological images and incomplete exams [11].

The Arnold-Chiari malformation type 1 was first found and reported on a 17-year-old girl who had hydrocephalus and died of typhoid fever[1]. It was associated with intramedullary cystic cavities (syringomyelia[8]) or scoliosis. In this type of malformation, there is a caudal displacement of the cerebellar tonsils that penetrate the spinal canal through the foramen magnum, extending until the atlas or axis[8], in addition to the medial parts of the lower lobes of the cerebellum forming conical projections and accompanying the spinal cord in the spinal canal[12].

The Arnold-Chiari malformation type 2 was first described in 1883 by the Scottish physician John Cleland, and referred as Basilar Impression Syndrome[1], characterized by a caudal displacement below the occipital foramen, the tonsils, the brainstem, the fourth ventricle and the vermis. It can coexist with other malformations; the most common is the spina bifida [12].

In the type 3, a complete herniation of the cerebellum is present in the spinal canal[12]. In type 4, there is a cerebellar hypoplasia[12], and this is a very rare form[8].

Other Arnold-Chiari malformations are types 0 and 1.5. Type 0 is characterized by minimal or absent cerebral hernia. Type 1.5 is characterized by tonsillar hernia without elongation of the brainstem or deformity of the fourth ventricle[12].

On May 6, 1916, van Chiari died of throat infection. He published around 180 papers between 1876 to 1916, and always cared to recognize the findings of other colleagues[1]. The findings about the Arnold-Chiari malformations were among his most important works. Besides van Chiari findings and Arnold contribution, other collaborations were made in the cardiovascular area, the approach to pituitary adenomas and on the Budd-Chiari Syndrome. In 1845, Budd described 5 cases of thrombosis of the hepatic vein; in 1899, Chiari made the first pathologic description of a liver affected by disease. Chiari was an illustrious physician, with many contributions to the scientific Medicine knowledge [8,11,12].

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