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Case Report

Chiari Network in Patient with Interatrial Septal Defect and Pulmonary Hypertension

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Abstract
Chiari network is a moving fibrous mesh-like structure that is visualized in 2% of cases in the right atrium. It is attached on one side to the valve of the inferior vena cava and on the other to different parts of the right atrium. In most cases this structure has no clinical significance and may be an accidental finding during echo- imaging. But sometimes it can be a source of thrombosis and in the future – the cause of thromboembolism, or, conversely, be an obstacle to the movement of thrombus.

Keywords
Chiari Network; atrial septal defect; pulmonary hypertension; right atrium; sinus venosus

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Background

The Chiari network was first described in 1875 when Carl Rokitansky found one case of network structure in the right atrium, but it was named after Chiari, embryologist and pathologist, who first described several cases of a mesh membrane located above the entry of the inferior vena cava. In 1879, in Prague, Chiari described the case of Rokitansky in his publication, as well as several other similar cases of a mesh membrane located above the entry of the inferior vena cava. [1]. Hans Chiari described abnormal fibrous structures extending from the edge of the inferior vena cava or coronary sinus valves. This mesh results from incomplete resorption of the right valve of the venous sinus. The other, the left venous sinus, preferably fuses with the atrial septum. [2]

The Chiari network is a movable, fibrous, filamentous structure that is visualized in the right atrium, attached to the Eustachian valve or other atrial structures. This element is an embryological remnant of the right valve of a venous sinus that appears at its incomplete resorption and mainly has no clinical value. The distribution of the Chiari network doesn't depend on age and gender. It is more common in people with other congenital anomalies, such as an open oval window and aneurysm of the atrial septum. It should be noted that sometimes the Chiari network can be the cause of thrombus formation, paradoxical embolism, obstruction of the pulmonary vessels, and tumor development. Cases of infectious endocarditis have also been described. [3, 4]. However, according to some authors, Chiari network, on the contrary, can serve as a natural filter that prevents massive pulmonary embolism through blood filtration. [5]

There may be various options for attaching the Chiari network. On the one hand, it can be attached to the Eustachian valve of the inferior vena cava, on the other – to different departments of the right atrium: to the valve of the main venous collector, which flows into the right atrium, the coronary sinus, to the wall of the atrium itself. [1]

The development of visualization methods of research makes it possible to detect the Chiari network as an accidental finding or in connection with any pathological conditions in 2% of cases. The detection rate of Chiari network during morphological studies is 1.3 - 4.0%. [6, 7]. The Chiari network has some diagnostic difficulties, including differential diagnosis, as it can be interpreted as vegetation, blood clots, damage to the tricuspid valve and even tumors.

1. Case report

Due to the rare detection of the Chiari network, we present our own case of detection of this formation. Patient D., aged 80 years, was treated in the osteo-purulent surgical department for osteomyelitis of the right thigh. Surgical intervention on the osteomyelitis of the femur was planned. The anamnesis of the disease showed that in 2016 the woman was performed endoprosthesis replacement of the right hip joint after fracture of the femoral neck. A year after the operation, pus began to discharge in the area of the prosthesis, a fistula has formed.

Life anamnesis has shown that the patient in the past very rarely sought the doctor's assistance, but she remembers that being a young woman it was difficult to perform physical activities, including sports. During examination pathological changes in the heart there were not detected. At the age of 70, type II diabetes was first detected. 5 years ago there were signs of cardiac arrhythmias – atrial fibrillation, permanent form. She was repeatedly treated as in-patient because of the appearance of edema on the lower extremities. Physical examination data: general condition is satisfactory, consciousness is clear, skin is pale, subcutaneous-fat tissue is developed moderately. Thoracic cage is normal, both halves participate in the act of breathing, there are no subcutaneous emphysema and crepitus. There is no shortness of breath. Respiratory rate is 16/min. Auscultation – vesicular respiration, no additional respiratory noise during examination is detected. There is no visible pulsation of the arteries, veins. The apex impulse is normal. Heart tones are of slightly diminished volume, arrhythmic melody, there is II tone accent over a.pulmonalis, systolic noise is heard at the apex of the heart. Heart rate is 85/in, Pulse 76/min, arrhythmic, pulse deficiency is 9, blood pressure is 140/80 mm Hg. The abdomen is soft, painless, symmetrical. The edge of the liver is felt 3-4 cm below the costal arch, painless, dense. The spleen is not palpated. Lower legs are cyanotic, with numerous tiny moist trophic ulcers. Laboratory-instrumental examination data: General blood test: erythrocytes 3.0×10^{12}/ l, hemoglobin 87 g/l, color index 0.75, anisocytosis is slightly pronounced, some erythrocytes are hypochromic, reticulocytes 0.3%, hematocrit 0.3, ESR 25 mm/h, leukocytes 5.6×10^{9}/l (segmentonuclear neutrophils – 72%, band forms 1%, immature 0%, eosinophils 1%, basophils 1%, lymphocytes 21%, monocytes3%), platelets 220×10^{9}/l. Glucose: 7.0 mmol/l. Coagulogram: Prothrombin time 14s, MNO – 1.1, ACTH 30s, activated recalcification time 82, plasma fibrin 3.9, prothrombin index 115%, thrombin time – 18s, soluble fibrin-monomer complexes – 0.4units, antithrombin III – 95%, fibrinogen – 3.4 g. General urine analysis: color – straw-yellow, transparent, density 1018g/l, protein 0.015g/l, glucose is absent, ketone bodies are absent, erythrocytes 1-2 in field of vision, leukocytes up to 4 in field of vision, cylinders are absent, salts are absent, bacteria are absent. ECG data: Atrial fibrillation, permanent form, tachy-systolic option. Vertical position of the electrical axis of the heart. Incomplete blockage of the right leg of the bundle of His. Left ventricular hypertrophy.

CHA2DS2-VASc - 7 points, EHRA - III.

Echo-CG data: Atrial and right ventricle dilation. Hypertrophy of both ventricles. Pulmonary hypertension – II degree (45 mmHg) Dilation of the pulmonary artery. Aortosclerosis. The overall contractility of the left ventricle is satisfactory, of the right one is reduced. Intracavitary formation: Chiari network is in the right atrial cavity. Pericardium: Separation of the leaves of the pericardium up to 5 mm. Aneurysm of the atrial septum in the area of the oval window. Secondary atrial septal defect up to 5 mm. Left-right shunt.

Treatment of the patient included the prescription of antiarrhythmic drugs, diuretics, antithrom-

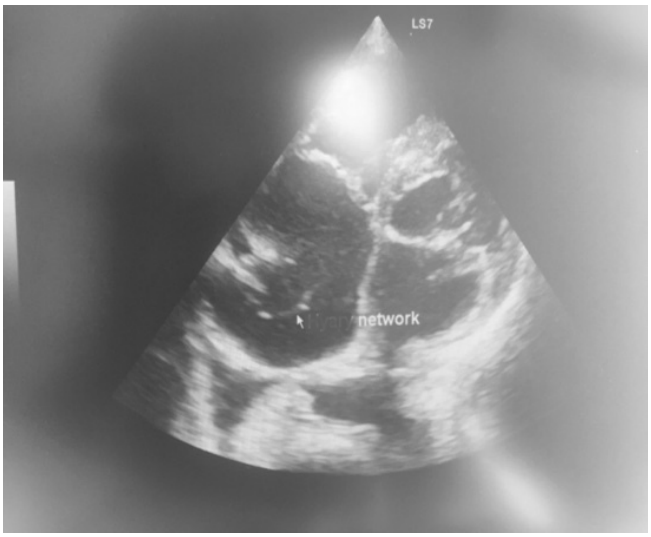

Figure 1. Echo CG of the patient D., 80 years. In the cavity of the right atrium, a Chiari network is visualized (in an arrow).

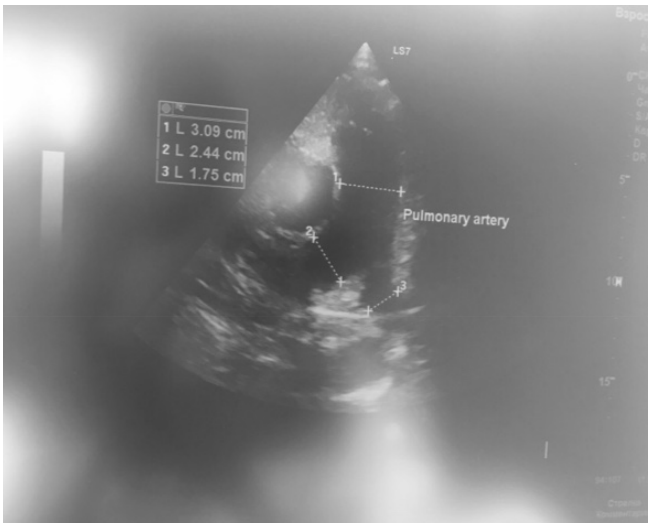

Figure 2. Echocardiography of the patient D., aged 80. Pulmonary artery enlargement is visualized.

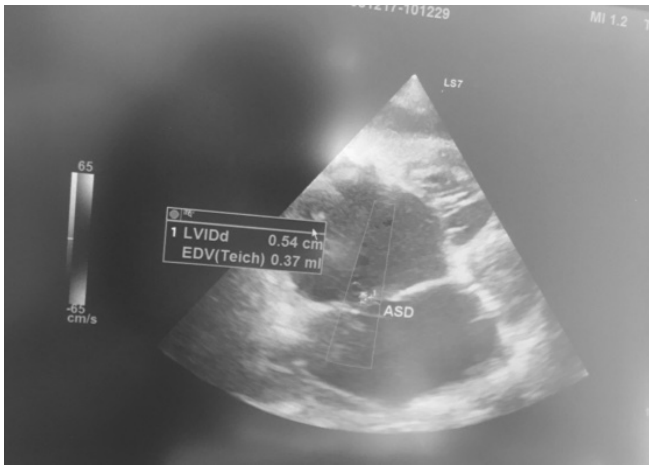

Figure 3. Echo CG of the patient D., 80 years. In the middle third of the interventricular septum (the area of the oval window) a tissue defect of 0.54 cm in size is visualized, with the signs of blood discharge from left to right – secondary defect of the interventricular septum.

botic therapy, ACE inhibitors, metabolically active medicines. Against the background of treatment, the patient's condition has improved. The patient was being prepared for surgical treatment of osteomyelitis.

2. Conclusions

A rare clinical case of primary Chiari network detection, combined with secondary atrial septal defect and pulmonary hypertension, is described. There is an interesting fact that over a long period of life the patient repeatedly sought the doctors' help, including cardiologists, but this formation has never been diagnosed before. There were no abnormalities in the development of the atrial septum; pulmonary hypertension, whose clinical features, in addition to the accentuation of the 2^{nd} tone on the pulmonary artery, was also absent.

Informed Consent

Written informed consent was obtained from the patient who participated in this case.

Conflict of Interest

The authors stated no conflict of interest.

Financial Disclosure

The authors declared no financial support.

References

[1] Alekhin MN, Dokina ED. Ekhokardiograficheskaya diagnostika seti Khiari. Ultrazvukovaya i funktsionalnaya diagnostika. 2014, 5: 119-127.

[2] Jansirani D Devi, Deep S Shiva, Anandaraja S. Anatomical Study of Chiari Network and the Remnant of Left Venous Valve in the Interior of Right Atrium // US National Library of Medicine National Institutes of Health. Hindawi Publishing Corporation Anatomy Research International Volume. 2015; 5 pages. DOI: https://doi.org/10.1155/2015/247680 [PMid:26442159 PMCid:PMC4579302]

[3] Loukas M, Sullivan A, Tubbs RS, Weinhaus AJ, DerDerian T, Hanna M. Chiari' s network: review of the literature. Surg Radiol Anat. 2010; 32 (10): 895-901. DOI: https://doi.org/10.1007/s00276-010-0639-z [PMid:20198476]

[4] Latif F, Peyton M, Laszik Z, Sivaram CA. Infective endocarditis of a papillary fibroelastoma on Chiari network of right atrium: a case report. J. Am. Soc. Echocardiogr. 2008; 21 (2): 188.e3-188.e4. DOI: https://doi.org/10.1016/j.echo.2007.08.033 [PMid:17928193]

[5] Helwig FC. The frequency of anomalous reticula inthe right atrium of the human heart "Chiari network". Report of eight cases. Am. J. Pathol. 1932;(1): 73-79.

[6] Werner JA, Cheitlin MD, Gross BW, Speck SM, Ivey TD. Echocardiographic appearance of the Chiari network: differentiation from right-heart pathology. Circulation. 1981; (5) 1: 104-109. DOI: https://doi.org/10.1161/01.CIR.63.5.1104 [PMid:7471371]

[7] Schneider B, Hofmann T, Justen MH, Meinertz T. Chiari' s network: normal anatomic variantor risk factor for arterial embolic events. J. Am.Coll. Cardiol.1995; 26 (1): 203-210. DOI: https://doi.org/10.1016/0735-1097(95)00144-O

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