

Case Report

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Unveiling the Chiari network: A case report of misdiagnosis mimicking pulmonary embolism and right atrium thrombus

Abstract

Background: The Chiari network consists of movable, mesh-like structures occasionally observed in the right atrium proximate to the openings of the inferior vena cava and coronary sinus.

Case Presentation: This case discusses a 75-year-old male initially suspected to have acute pulmonary thromboembolism, but we misdiagnosed the Chiari network as a thrombus in the right atrium. Subsequent echocardiography revealed a Chiari network in the right atrium. Accurate identification using transthoracic echocardiography, Doppler echocardiography, is crucial.

Conclusion: Accurate identification of the Chiari network is essential for optimal patient care. Thorough evaluation is crucial in cases suggestive of acute pulmonary thromboembolism or right atrium thrombus.

Keywords: Chiari network, Acute pulmonary thromboembolism, Doppler echocardiography, right atrial net.

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The Chiari network is a mesh-like, movable structure located within the right atrium, situated near the entrance of both the inferior vena cava and coronary sinus, which represent congenital remnants of the right valve of the sinus venosus, resulting from incomplete resorption of this structure during embryonic development (1). Some studies suggesting its potential association with thromboembolism, infective endocarditis, and supraventricular tachyarrhythmias (1, 2). Sory et al. suggest the term "right atrial net" as an alternative to "Chiari net," referring to anomalies that encompass the valves of the inferior vena cava and coronary sinus, as well as strands within the right atrium linking these valves to the crista terminalis, right atrial wall, or interatrial septum (2). Various studies have reported the prevalence of the Chiari network to range from 2% to 13.6% (2) or 1.3% to 4% founds in autopsy studies (1, 3, 4).

It is crucial to employ appropriate imaging techniques to distinguish this phenomenon from other right atrial abnormalities, such as right heart vegetation, flail tricuspid leaflet, ruptured chordae tendineae of the tricuspid valve, and right heart mass (thrombus) (5). Real-time 2D transthoracic Doppler echocardiography is a useful diagnostic tool for making a definitive diagnosis of a Chiari network and differentiating it from other structures in the right atrium, such as the Eustachian and Thebesian valves (6). Likewise, MRI imaging can assist in characterizing and distinguishing true cardiac masses from anomalous cardiac formations like Chiari network (7). The extension of the Chiari network into the right ventricle can result in notable tricuspid regurgitation (8).

In this paper, we detailed a case involving a substantial Chiari network, which resembled a right atrial thrombus in a patient diagnosed with dilated cardiomyopathy, accompanied by an enlargement of the left cardiac cavity.



Case Presentation

On September 5th, 2016 (09.35 PM) a 75-year-old man was admitted to our Clinical Hospital “D-r Trifun Panovski” – Bitola, in the Department of Cardiology and coronary disease, presenting a constellation of distressing symptoms. He described an overwhelming sensation of breathlessness, chest pain, accompanied by dyspnea, shortness of breath, a profound sense of fatigue, and palpitations that seemed to reverberate through his chest. Additionally, he recounted experiencing a clammy sheen of cold sweat across his forehead and chest, an unsettling physical manifestation of his discomfort. Amidst these troubling sensations, he endured bouts of severe chest pain, each throb a sharp reminder of the urgency of his condition. His symptoms started 2 hours ago, and then he felt that his heart was beating very fast.

The patient has a medical history significant for several chronic conditions. He was diagnosed with Chronic Obstructive Pulmonary Disease (COPD) 18 years ago, hypertension 14 years ago, and has been undergoing treatment for hyperlipidemia for the past 8 years. His medication regimen includes regular use of beta blockers, occasional use of aminophylline, and various antihypertensive drugs. In terms of lifestyle factors, the patient consumes up to 1 unit of alcohol per day and has a significant smoking history of 68 pack-years. It is important to note that he has never experienced symptoms such as abdominal burning or swelling and has no history of gastritis. Upon admission, his pulse was recorded at 130/min and blood pressure at 135/90mmHg. The initial laboratory results and their changes during his hospital stay are detailed in table 1.

Table 1. Variation of clinical parameters during hospitalization

Clinical parameters	05 Sept	07 Sept	09 Sept	12 Sept
RBC (x 10 ⁶ /L)	5.48	5.29	5.52	5.47
HGB (g/L)	154.0	151	158.0	155.0
PLT (x 10 ³ /L)	289.0	276	309.0	297.0
WBC (x 10 ³ /L)	13.3	9.7	10.6	9.1
Glikemia (mmol/L)	6.7	8.32	7.1	6.9
ALT (U/L)	29.0	36.0	27.0	31.0
AST (U/L)	187.0	129.0	43.0	40.0
CK (U/L)	1856.0	865.0	416.0	120.0
CK-MB (U/L)	416.41	85.1	43.1	21.6
LDH (U/L)	930.0	1093.0	765.0	497.0
Urea (mmol/L)	10.5	10.2	9.7	8.6
Creatinin (μmol/L)	115.0	118.3	112.1	110.7
Total Bilirubin (μmol/L)	16.5	17.2	15.9	16.9
Direct Bilirubin (μmol/L)	6.5	6.8	6.1	6.7
Cholesterol (mmol/L)	6.83	6.72	6.81	6.62
HDL-Cholesterol (mmol/L)	1.97	1.95	1.93	1.85
LDL-Cholesterol (mmol/L)	4.4	4.38	4.43	4.41
Triglycerides (mmol/L)	0.89	0.78	0.90	0.72
Troponin T (ng/L) (more than >)	3000	2000	2000	2000
D-Dimer (mg/mL)	2.74	1.93	1.14	0.54

RBC, red blood cells; HGB, hemoglobin; PLT, platelets; WBC, white blood cells; ALT, alanine aminotransferase; AST, aspartate aminotransferase; CK, creatine kinase; CK-MB, creatine kinase - miocardial band; HDL, high density lipoprotein; LDL, low density lipoprotein.

On admission, the patient's chest x-ray (figure 1) revealed bilateral hiliobasal pulmonary stasis and chronic peribronchitis, with a zone of reduced transparency on the left lung and marked left heart enlargement. The cardiothoracic ratio was 0.593 without pleural effusion. The electrocardiogram (ECG), conducted with a 12-lead, showed atrial fibrillation with rapid ventricular response and left bundle branch block, a ventricular rate of 126 bpm, a QRS duration of 174 ms, and QT/QTc intervals of 408/480 ms.

Gray scale and Doppler echocardiography:

Echocardiographic examinations were performed using a GE HealthCare Voluson P8 ultrasound machine (General Electric Company, Boston, Massachusetts, USA 2010) equipped with a 3Sc-Rs cardiac Doppler probe (Phased Cardiac Ultrasound Transducer, resonant frequency of 1.3 to 4.0 MHz).

There are signs of dilated cardiomyopathy with enlargement of the left cardiac cavity above all, decreased systolic function of the left ventricle (ejection fraction, EF 20%) which has a significant disturbances of the kinetics – akinesia of apical third of the front wall, all posterior wall, mid and apical third if the inferior wall and top. There is hyper contraction - compensatory of lateral wall and part of lateral wall. There are signs of small shock volume. In the enlarged left cavity no foreign mass is visible. Mitral regurgitation (MR 2-3+) of muscular origin is registered. Right heart is border-line bigger. In the right atrium a

floating mass 1x1 cm is registered in the level of vena cava inferior (IVC). It could be a thrombus but the lung artery is varicose. There is tricuspid regurgitation (TR1+) and christa terminalis (Chiari network). Pericard is with no visible changes. Dilated aorta is visualized in the exit part (4.0 cm) and in remaining segments normal width.

Using the Color Doppler flow mapping technique, two independent observers identified persistent turbulence in early diastole within the right atrium. The second flow, characterized by a retrograde blue mosaic signal due to a turbulent aliased jet, extended from the mitral valve into the right atrium, away from the transducer. These findings were observed in the presence of a Chiari network with a floating mass in the right atrium. 2D Doppler echocardiography images are included in figures 2 and 3.

Throughout his 8-day hospital stay, the patient received a regimen comprising cardiotonics (Cedilanid), diuretics (Furosemid), beta blockers (Nebivolol), anticoagulants (Enoxaparin), hepatoprotective therapy, l-ornithine l-aspartate (Hepa-Merz) and opioid pain killer (Tramadol), resulting in both subjective and objective improvement. Upon discharge, he was counseled and prescribed a treatment plan incorporating beta blockers, antihypertensives, diuretics, and anticoagulants, with a follow-up appointment scheduled in 7 days. The participant in this case report signed an informed consent and this case presentation was approved by Ethics Committee of our institution (code 01.12.2016/2cr).

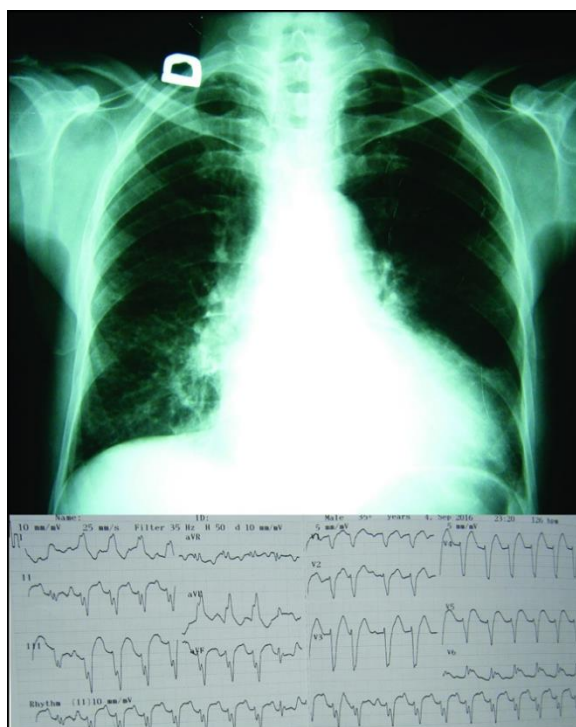


Figure 1. Electrocardiogram and Chest X-ray radiograph of the patient at hospital admission showed bilateral hiliobasal pulmonary stasis with a zone of left reduced lung transparency

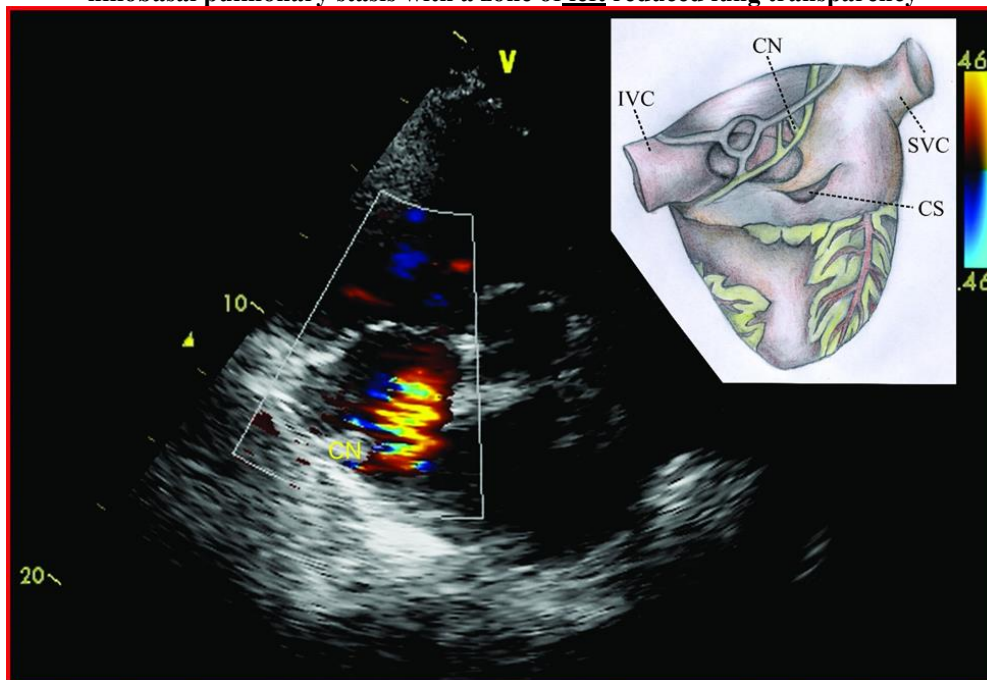


Figure 2. Live real time two-dimensional trans-thoracic Doppler echocardiography showing the Chiari network (CN) in right atrium presented as a floating mass. Hand-drawn picture illustrating the Chiari network within the right atrium (displaying the openings of the inferior vena cava, coronary sinus, and the Chiari network). Abbreviations used include IVC for inferior vena cava, CN for Chiari network, and SVC for superior vena cava, and CS for coronary sinus.



Figure 3. Echocardiography showing the Chiari network (CN) in right atrium presented as a floating mass (9.7 x 10.1 mm)

Discussion

Chiari's networks are described as delicate, thread-like strands that link the inferior vena cava and coronary sinus valves to the crista terminalis, or as sieve-like fenestrations within these valves (1, 2), for first time documented in 1897 by Hans Chiari, an Austrian pathologist in 13 human right atria (1) presented as a reticulated network of fibers within the right atrium which included the inferior vena cava and the valve of the coronary sinus (4). With an estimated

occurrence rate of around 2% and uncommon connections to pathology, Chiari network may be incidentally detected during diagnostic imaging procedures or surgical interventions (2). Initially, our diagnostic suspicion centered on this condition (thrombus) until a 2D Doppler color echocardiography confirmed the accurate diagnosis of Chiari network. I want to emphasize that atrial arrhythmia was present in our case, and it was connected to the Chiari network that we identified. Benbow et al. (5) proposed that

the existence of myocardial fibers within the reticulated threads of the network might predispose to arrhythmias. In Roldán et al.'s investigation (8) half of the ten patients with Chiari network in their right atrium and no other cardiac issues were found to have supraventricular arrhythmias (9). This observation led to the speculation that the presence of Chiari network might be linked to such arrhythmias. Our patient presented with symptoms commonly associated with acute pulmonary thromboembolism (APTE), including chest pain, dyspnea, and elevated levels of cardiac troponin T and D-dimer (10). These symptoms, along with the laboratory findings of elevated troponin T and elevated D-dimer, led to the initial suspicion of pulmonary thromboembolism (11).

However, upon further investigation (ECG, 2D Doppler echocardiography) it was determined that the patient did not have a pulmonary embolism, but Chiari network. This highlights the importance of thorough evaluation and differential diagnosis in cases presenting with symptoms suggestive of APTE to avoid misdiagnosis and ensure appropriate treatment. Another diagnosis represents a stumbling block in the differential diagnosis of Chiari network, which is a thrombus in the right atrium. The first B-mode echocardiography detect a false suspected diagnosis of a thrombus in the right atrium, which was denied by the repeated 2 D Doppler echocardiography examination. The diagnosis of Chiari network by Doppler echocardiography excluded the possibility of a thrombus in the right atrium.

Several studies found an increased prevalence of Chiari malformations among individuals with neurofibromatosis (13) and Behcet disease (14) compared to the general population. In Erdogan et al. (2017), a highly mobile structure near the inferior vena cava and patent foramen oval was seen on echocardiography (15). During a planned aortocoronary bypass surgery, a net-like structure was found at the opening of the inferior vena cava and coronary sinus, confirmed to be the Chiari network and excised. There was no thrombus, and tricuspid valve incompetence was ruled out (15). In another study involving an 82-year-old patient, a mobile echogenic mass in the left atrium was detected by transesophageal echocardiography. This study reported a unique and rare case of the Chiari network herniating into the left atrium following percutaneous interventions requiring transseptal puncture. The initial suspicion of a thrombus in the left atrium was ultimately rejected (16).

To ensure accurate diagnosis of the Chiari network, a comprehensive approach is essential, utilizing transthoracic

2D Doppler transthoracic echocardiography. Multiple transthoracic windows and views such as the apical 4-chamber view, parasternal right ventricular inflow tract view, and a basally angulated parasternal short axis view must be employed for thorough assessment of the right atrium, ensuring meticulous evaluation and precise diagnosis (6). Advanced echocardiography using Color Doppler is essential when a thrombus is suspected but not clearly identifiable by basic methods, such as B-mode echocardiography. This technique is particularly valuable for identifying intracardiac thrombi in patients with atrial fibrillation (as in our patient), assessing embolic risk, and differentiating between thrombi and anatomical structures like the Chiari network in the right atrium. Its superior diagnostic accuracy and detailed hemodynamic information make Color Doppler echocardiography crucial for comprehensive cardiac evaluation. Limited experience in echocardiography using b-mode ultrasonography, coupled with misinterpretation of elevated cardiac troponins and D-dimers, resulted in erroneous diagnoses of right atrial thrombus and APTE.

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Conflict of interests: The authors declare that there is no conflict of interest regarding the publication of this paper.

Ethical approval: Obtained from the Ethics Committee of Clinical Hospital – Bitola (code 01.12.2016/2CR); patient provided written informed consent.

Authors' contribution: Petar Avramovski, developed the initial concept and design of this case report. Being the corresponding author, he participated in writing and submitting the paper, organizing the documentation, processing the text and images, wrote the background section of the manuscript. He coordinated patient recruitment, consent, and data collection and verified the accuracy and consistency of collected data. Zaklina Servini, admitted the patient to the hospital and, as a cardiologist, was responsible for his therapy and treatment. She performed the initial B-mode echocardiographic examination, documented the anamnesis and medical history, raised suspicions that led to a repeated

echocardiographic examination, and gathered patient data, clinical information, and imaging results. Maja Avramovska reviewed the manuscript for language accuracy and grammatical correctness, prepared and formatted tables and figures for case report and managed and formatted references according to journal guidelines. She was active in submitting documentation to the ethics committee and obtaining consent. Kosta Sotiroski, an experienced professor of statistics, compared our laboratory results, recommended relevant references, contributed to creating the Excel tables, participated in writing the discussion, and provided statistical interpretations of the results. Additionally, he offered critical feedback and suggestions for improving the manuscript. Aleksandra Arandjelovic, as a professor of ultrasonography and interventional cardiology, she performed the second echocardiographic examination and established the diagnosis. She provided detailed B-mode and Doppler echocardiographic descriptions for the medical reports and initiated the proposal to publish this case in a scientific journal. Additionally, she reviewed the manuscript for natural language flow and readability.

Aleksandra Servini assisted with writing and revising the case report, contributed to specific sections of the manuscript, such as the case presentation and conclusion and provided technical support for medical imaging equipment. Vesna Siklovska an experienced radiologist, actively participated in radiographic recording and interpretation of x-ray images, creator of the text explaining the images of imaging techniques, designed visual content and enhanced images using graphic design software, prepared and formatted figures for the case report. Stefan Talev wrote and reviews the initial concept and study design, reviewed the manuscript for language accuracy and grammatical correctness, and reviewed the manuscript for natural language flow. He compiled and organized laboratory test results relevant to the case. Irena Trajcevska provided technical support for medical imaging equipment, gathered patient data, and imaging results and drafted the main sections of the manuscript. She actively participated in the radiographic recording and interpretation of the x-ray images as a second radiologist, independently and blindly from the interpretation of the first radiologist.

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