

 Mehmet Ali Sahin,  Erkan Kuralay

Alife Hospital, Department of Cardiovascular Surgery,
Ankara, Türkiye

Received: 02 May, 2024
Accepted: 10 July, 2024
Published: 12 July, 2024

Corresponding Author: Erkan Kuralay
Alife Hospital, Department of Cardiovascular Surgery,
Ankara, Türkiye
Email: erkankuralay@gmail.com

CITATION

Sahin MA, Kuralay E. Chiari network with interatrial septum defect and aneurysm. AZJCVS. 2024;5(2):51-3.

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INTRODUCTION

The embryonic right valve of the sinus venosus protrudes into the right atrium towards the foramen ovale to direct the oxygen-rich blood from the inferior vena cava across the atrial septum to supply the left heart and upper body. This valve tissue regresses during later growth leaving remnants in the right atrium. Usually, these are seen as a haemodynamically insignificant Eustachian valve adjacent to the orifice of the inferior vena cava and a much smaller Thebesian valve next to the orifice of the coronary sinus on the atrial septum. Infrequently, with fragmentary resorption of the embryonic valve membrane, a network of interconnected filamentous strands can result which reaches from the inferior vena cava across the right atrium to the atrial septum and in some can elongate or stretch. Right valve is more likely to be elongated postnatally, leading to the formation of a Chiari network (CN). A CN is characterized by whip-like motion within the right atrium and attachment to the wall of the right atrium in close proximity to the entrance of the inferior vena cava. In patients undergoing transesophageal echocardiography, the prevalence of Chiari's network was 2%, as in autopsies (1). On microscopic analysis in

children and adults, the CN has been found to consist of strands of cardiac muscle surrounded by elastic tissue. Cases have been reported of a prominent prolapsing CN discovered as an incidental echocardiographic finding without other consequence. However, there are also many case reports of this condition being associated with arrhythmias, atrial septal defects, interatrial septum aneurysm (IASA), tricuspid valve regurgitation, and significant obstruction to right ventricular outflow tract flow. Although it is generally benign, the persistence of this remnant has been found to be associated with cardiovascular complications such as supraventricular arrhythmias, interatrial septal defects, thromboembolism or cyanosis (2). Herein: we present adult patients with CN.

CASE REPORT

Fifty-five years old male patients was admitted. Patients suffered transient ischemic attack with right side hemiparalysis nine months ago. Neurological deficit spontaneously recovered.

Patient was on normal sinus rhythm and incomplete right bundle block pattern was found in preoperative electrocardiogram.

Chiari network with interatrial septum defect and aneurysm

Abstract

Chiari network is a embryogenic remnant of sinus venosus. This network tissue regresses during later growth leaving remnants in the right atrium. Chiari network does not regress completely and remnants remain in the right atrium in 2% of population. Remnants of Chiari-network are frequently confused with thrombus and vegetation on echocardiographic examinations. Chiari network is usually associated with interatrial septal defect and interatrial septal aneurysm. Chiari network can cause tricuspid valve regurgitation, atrial fibrillation, paradoxical embolism and bacterial embolism. We have performed surgery for Chiari network which was caused transient emboli and transient atrial fibrillation.

Keywords: Chiari-network, transient embolic attack, atrial fibrillation

Patient was also complained palpitation attacks. CN was accidentally found in routine transthoracic echocardiographic examination. Transesophageal examination has revealed interatrial septal aneurysm and CN (Figure 1). Saline test revealed bubble transition to the left atrium. Patient was taken into operation room. Median sternotomy was done. Pericardium harvested and processed with glutaraldehyde solution. Ascending aorta and bi-caval cannulations were done. Ascending aorta cross-clamped. Right atriotomy was done with standart fashion. Whip-like CN tissue origination was completely resected from inferior cava orifice (Figure 2. A and B). IASA was observed. There were two different interatrial septum defect found. IASA was completely removed. Defect was closed with glutaraldehyde processed pericardium and right atrium was closed. Postoperative course was uneventful. Patient discharged at 3th postoperative days. Patient was completely asymptomatic after surgery. There was not any supraventricular tachyarrhythmias during the follow-up.

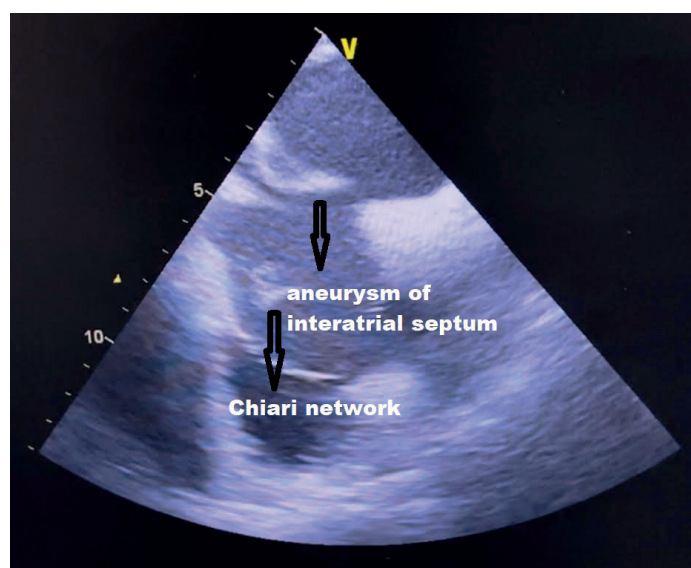


Figure 1. Intra-operative tee view: Whip-like Chiari network in the right atrium and aneurysm of interatrial septum are clearly demonstrated

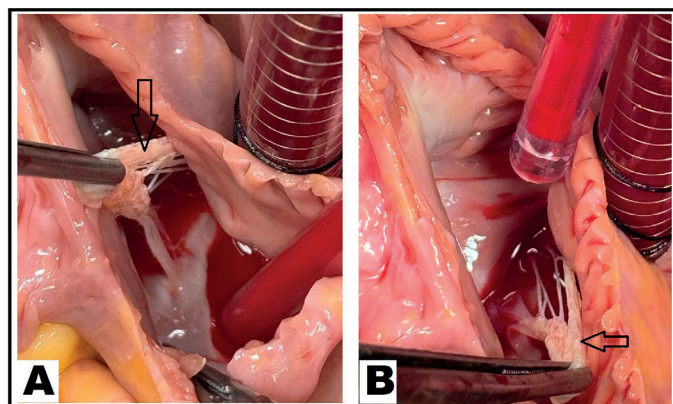


Figure 2. Surgical view: **A.** Chiari network retracted to leftside and chiari's attachments are seen. **B.** Chiari network retracted to rightside and chiari's attachments around the vena cava inferior orifice are demonstrated

DISCUSSION

CN generally associated with atrial septal defect (80%) and IASA (20%). Generally Chiari Network may confused with valve disruption, vegetation, thrombus at initial echocardiographic examination. CN maintains an embryonic right atrial flow pattern into adult life and directing the blood from the inferior vena cava toward the interatrial septum. CN may facilitate paradoxal embolism and right-side endocarditis in patients with atrial septal aneurysm, patent foramen ovale. Schneider B (3) have reported systemic arterial embolic events. CN is not completely benign entity due to above mentioned complication. The CN is considered unusually prominent when large enough to prolapse through the tricuspid annulus into the right ventricle during diastole (4) and may cause tricuspid regurgitation (5). CN may interfere right-side catheterization and pacemaker lead insertion. Web-like CN also prevent pulmonary embolism due to blockage of thrombus around the inferior vena cava orifice (6).

Management of the CN and patent foramen ovale and interatrial septal aneurysm coexistence in asymptomatic and symptomatic patients is still contravertial. A prophylactic anticoagulant or an antiplatelet is not indicated in patients who did not have a history of transient ischemic attack (TIA) or cerebrovascular accident. However, it is a well-recognized issue that an aneurysmal interatrial septum increases the risk of paradoxical embolism from a patent foramen ovale (PFO). Hence, in the case of venous thromboemboli (VTE) in a patient who already has had a PFO and aneurysmal interatrial septum (IAS), there would be a significant risk for left circulation embolism. Since a small deep vein thrombosis may cause a catastrophic cerebrovascular accident (CVA), physicians must be cautious for VTE detection in such high-risk patients. In the left circulation thromboembolism such as CVA and TIA, if a patient has a PFO, it must be classified as a PFO-related embolism instead of a cryptogenic embolism. However, together with the treatment, other causes of embolisms such as atrial fibrillation (AF) rhythm should be evaluated. The novel criteria of risk of Paradoxical Embolism (RoPE score) could determine whether a PFO has played a role in CVA. Treatment methods of PFO-related CVA are controversial. Some guidelines advise using antiplatelet agents for secondary prevention, but the newer approaches mostly insist on the superiority of oral anticoagulants. Considering secondary prevention, PFO closure has not been well-established as an absolute effective procedure, but it must be considered in high-risk patients. In addition, a PFO closure might prevent AF rhythm from happening which is another cause of the left circulatory embolism. We have performed surgery due to TIA history and atrial fibrillation attacks in our patients.

CONCLUSION

A CN is characterized by whip-like motion within the right atrium and attachment to the wall of the right atrium in close proximity to the entrance of the inferior vena cava. It may confused with right atrial mass, thrombus, vegetation. CN may

cause severe supraventricular tachycardia attacks and cerebral emboli. Surgical intervention is required complicated CN cases.

Conflict of Interests: The authors declare that there are no conflict of interests.

Financial Disclosure: The authors declare that there is no financial support.

Patient Informed Consent: Informed consent was obtained from the patient. Signed form is included the patient file.

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