Chiari network: A case report and brief overview

A.K.M. Monwarul Islam a,⇑, Lima Asrin Sayami a, Shahana Zaman a

a Department of Cardiology, National Institute of Cardiovascular Diseases, Dhaka
b Bangladesh

The Chiari network is mobile, net-like structures occasionally seen in right atrium near the opening of inferior vena cava and coronary sinus. This is usually of no clinical significance and is often diagnosed incidentally. However, sometimes it may cause diagnostic confusion with right atrial pathologies, and may favour thromboembolism by causing flow obstruction. It may be associated with infective endocarditis, arrhythmias, and migraine. Sometimes, it acts as a physical barrier during invasive procedures. The Chiari network has also been described to protect from pulmonary embolism by acting as an inferior vena cava filter due to its sieve-like effect at the cavo-atrial junction. Here, the Chiari network has been described in a case of Ebstein anomaly of tricuspid valve which produced diagnostic confusion during echocardiography. A brief overview has also been presented.

© 2013 King Saud University. Production and hosting by Elsevier B.V. All rights reserved.

Keywords: Coronary sinus, Inferior vena cava, Ebstein anomaly

Introduction

The Chiari network, encountered infrequently in the right atrium, is a fenestrated, net-like embryonic remnants of valves of sinus venosus, lying closely in relation to the inferior vena cava and coronary sinus, sometimes connecting these with other right atrial structures [1]. It is usually of no clinical significance. However, it may at times produce diagnostic confusion, cause thromboembolism, infective endocarditis, arrhythmias, or cause physical barrier to invasive procedures [1]. Here, the Chiari network has been described in a case of Ebstein anomaly of tricuspid valve which produced diagnostic confusion during echocardiography.

Case presentation

A 19-year-old lady with Ebstein anomaly of tricuspid valve, presented to the coronary care unit with palpitation and breathlessness for 2 h. Her pulse was 160/min, regular, and blood pressure 90/50 mmHg. Electrocardiography (ECG) showed regular wide-complex tachycardia (Fig. 1). Echocardiography revealed apical-wards shifting of tricuspid valve, atrialization of right ventricle and a secundum-type atrial septal defect with left-to-right shunt. The junior doctors were confused to see whip-like structures extending from the opening of inferior vena cava (IVC) and moving freely within the right atrial cavity (Fig. 2). Presence of
The Chiari network was ascertained. The wide-complex tachycardia was controlled with IV amiodarone. There was accessory atrioventricular conduction in sinus rhythm ECGs. Previous documents showed similar episodes of wide-complex tachycardia responding to IV verapamil. She was kept on oral amiodarone keeping in mind the provision of cardiac cath, electrophysiological study and surgery.

Discussion

The Chiari network is a meshwork of thread-like strands connecting the edges of the inferior vena cava and coronary sinus valves with the crista terminalis, or sieve-like fenestrations in the valves [2]. Hans Chiari, an Austrian pathologist, described this for the first time in 13 human right atria in 1897 [3].

The Chiari network results from failure of resorption of the right sided sinus venous valve. Developmentally, the right valve of the sinus venosus evolves into two valves: the valve of the inferior vena cava (Eustachian valve) and the valve of the coronary sinus (Thebesian valve). During involution of these valves, the tissue undergoes fenestration so that a network may be formed from remnants that usually disappear [4–6] (Fig. 3). The prevalence of Chiari network has variably been reported to be 2–13.6% [2,7,8].

Echocardiography is an excellent tool for the diagnosis of the Chiari network. In transthoracic and transesophageal echocardiography, this network presents as a highly mobile, highly reflectant echo target in several locations in the right atrium [9]. Recently, real time 3D transthoracic echo has been found useful in making a definitive diagnosis of the Chiari network and differentiating it from other structures in the right atrium such as the Eustachian and Thebesian valves [10]. Newer imaging modalities, particularly, cardiac magnetic resonance play complimentary roles with echocardiography in differentiating Chiari network from right atrial pathologies [11].

The Chiari network is mostly an incidental finding, and itself bears no major clinical significance. However, sometimes it poses diagnostic difficulties during echocardiography where it could be
misdiagnosed as right atrial thrombi, tumors and vegetation [9,12,13]. It may produce continuous or systolic murmur, clinically, that could be confused with bruit de Roger of ventricular septal defect [14,15]. The Chiari network protruding into the right ventricle may produce significant tricuspid regurgitation [16]. It may favour persistence of a patent foramen ovale and formation of an atrial septal aneurysm and, may facilitate paradoxical embolism [8]. It may be associated with thrombi formation, and part of the strands may embolize [5,17,18]. Infective endocarditis has been reported in association with the Chiari network [13,19,20]. There may be abnormal atrial depolarization favouring supraventricular arrhythmia [21]. During invasive procedures, catheters [15,22,23], guidewires [24] and pacemaker leads [25,26] may get entrapped within the net. During retrograde cardioplegia before open-heart surgery, the Chiari network may cause difficulties in the cannulation of the coronary sinus [27].

Presence of the Chiari network sometimes may be confused with the diagnosis of cor triatriatum dexter or even Ebstein anomaly of the tricuspid valve [28–30]. Cor triatriatum dexter also results from incomplete resorption of the right sinus venosus valve, but the dividing septum of cor triatriatum dexter is much thicker with few or no
fenestrations. Unlike the Chiari network, it may cause obstruction to blood flow and is usually associated with other congenital abnormalities such as atrial septal defect and tricuspid atresia [28–30].

Interestingly, the Chiari network has been described to play a protective role in special situations, such as an inferior vena cava filter in patients with thrombophilia [31], and holding thrombus in the network preventing embolization [32,33]. This network has also been described in association with neurofibromatosis [34], Behçet syndrome [35], platypnea–orthodeoxia [36] and migraine [37].

In the case presented here, the Chiari network in probably an incidental finding. However, its presence might facilitate the occurrence of accessory pathway of atrioventricular conduction resulting in Wolf–Parkinson–White syndrome; otherwise, the accessory pathway might be a well-recognized association of Ebstein anomaly of tricuspid valve.

Conclusion

The Chiari network is an uncommon diagnosis. It has to be recognized appropriately, otherwise it may lead to misdiagnosis. This apparently ‘normal variant’ may not be always so. Respective clinicians should keep its presence in mind while doing echocardiography to avoid diagnostic dilemma, while performing cardiovascular interventions to overcome difficulties, and while dealing with patients to make the appropriate diagnosis.

Conflict of Interest

Authors have no conflict of interest to declare.

References


