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International Journal of Medical Case Reports and Medical Research

Case Report

Chiari Network Induced Tricuspid Regurgitation an incidental Finding on perioperative Trans- Esophageal Echocardiography in a Patient Undergoing Laparoscopic Cholecystectomy

Vishnu Datt*, DA,DNB, MNAMS, Priyanka Ahlawat, Diksha Datt Md St, Priyanka Dhahiya MD st.1, Pawan Yadav, Priyanka Kaushik MD st, Nithiya SR

Department of Anaesthesiology and Critical Care1, & PSM2,and surgery SGT medical College, Gurugram, Haryana

Article Info

Received: December 24, 2024
Accepted: December 27, 2024
Published: January 11, 2025

***Corresponding author:** Vishnu datt, Professor & HOD Anesthesiology,SGT Hospital, Gurugram, HaryanaFormer Director, Cardiac Anesthesia,G B Pant hospital, New Delhi.

Citation: Vishnu Datt, Priyanka Ahlawat, Diksha Datt, Priyanka Dhahiya, Pawan Yadav. (2025) "Chiari Network Induced Tricuspid Regurgitation an incidental Finding on perioperative Trans- Esophageal Echocardiography in a Patient Undergoing Laparoscopic Cholecystectomy.", International Journal of Medical Case Reports and Medical Research, 3(4); DOI: 10.61148/2994-6905/IJMCRMR/164.

Abstract:

It is known that some neurological conditions such as epilepsy and headache Chiari network is a remnant of eustachian valve located in the RA that helps divert oxygenated blood in a fetus from the inferior vena cava (IVC) towards the foramen ovale to the LA and LV and then to the aorta. The incomplete involution of the fetal sinus venosus valves results in redundant Chiari Network (CN). The CN is a rare anomaly and described as a web-like network in the RA that is usually thin and small and remains non-significant in most of the patients. However, it may be a risk factor for development of arrhythmias, thromboembolism, pulmonary arterial hypertension, endocarditis, atrial septal aneurysm, and transient ischemic attack and stroke due to paradoxical systemic embolism through the patent foramen ovale (PFO). Sometimes it can be long enough to protrude into the right ventricle during diastole with atrial contraction and affect the competency of the tricuspid valve during systole resulting in tricuspid regurgitation (TR). However, the extent of CN prolapse gradually decreases and many related abnormalities resolve during growth of the patient. We present an incidental diagnosis of CN induced moderate TR on perioperative transoesophageal echocardiography (TEE) evaluation in a 22 yrs old male patient undergoing laparoscopic cholecystectomy. The TEE was performed for the evaluation of structural cardiac defects of the ECG findings suggestive of RVH and prominent J waves.

Keywords: Chiari Network; Cholelithiasis; embolism; incidental finding PAH, RVH; Tricuspid Regurgitation

Introduction:

A Chiari network (CN) has been described as a filamentous, weblike structure in the right atrium that results from incomplete resorption of the embryonic sinus venosus. The prevalence of Chiari network varies from 2–13.6%. Usually, it has no clinical significance and often observed incidentally during perioperative transoesophageal echocardiography (TEE) evaluation of the patients undergoing various paediatric and adult cardiac surgical procedures. However, rarely it predisposes the patients to the development of arrhythmias, thromboembolism, pulmonary arterial hypertension, endocarditis, atrial septal aneurysm, tricuspid regurgitation (TR), and transient ischemic attack and stroke due to paradoxical systemic embolism through the patent foramen ovale(PFO). Here, the report on a chiari network induced TR has been described on the perioperative TEE evaluation in a 22 yrs male patient undergoing laparoscopic cholecystectomy. The perioperative, TEE was indicated to rule out the structural cardiac defect for RVH, prominent J wave and RBBB findings of ECG evaluation.

Case Report:

A 22 yrs male presented with acute pain on the right hypochondrium radiating to the back for one month. It was associated with indigestion, dyspepsia, belching and few episodes of vomiting. It was not associated with dyspnoea, chest pain, palpitation, sweating or syncope. USG of upper abdomen revealed multiple gall stones, with mild wall thickening and normal CBD. He was scheduled to undergo laparoscopic cholecystectomy after routine preoperative evaluation. On physical examination nothing was significant. His HR was 80bpm regular, systemic BP was 134/80 mmHg. His SGOT was 114U/L, SGPT 85U/L, and prothrombin time was 14.20sec, INR was 1.18. All other haematological and biochemical values were with in normal limits. Chest X-ray showed normal cardiac size and clear both lungs' fields, but the pulmonary arteries were prominent.

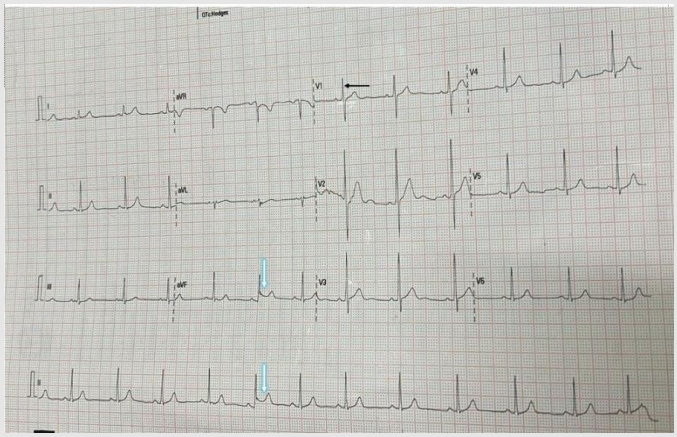

Figure 1: A standard 12 lead ECG reveals prominent J wave in Lead II and AVF suggestive of early repolarization in Brugada syndrome (white arrow), and prominent S wave in lead V1 suggestive of RVH (black arrow).
RVH- right ventricular hypertrophy

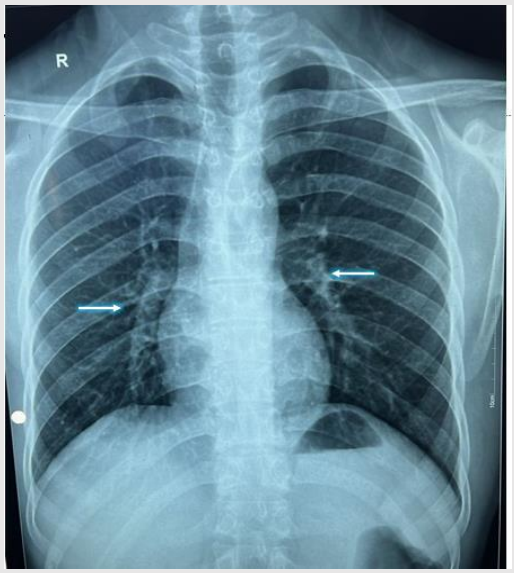

Figure 2: Chest X-Ray- PA view. shows normal cardiac size and clear both lungs' fields, but the pulmonary arteries are prominent suggestive of presence of PAH (arrows)
PAH- pulmonary arterial hypertension

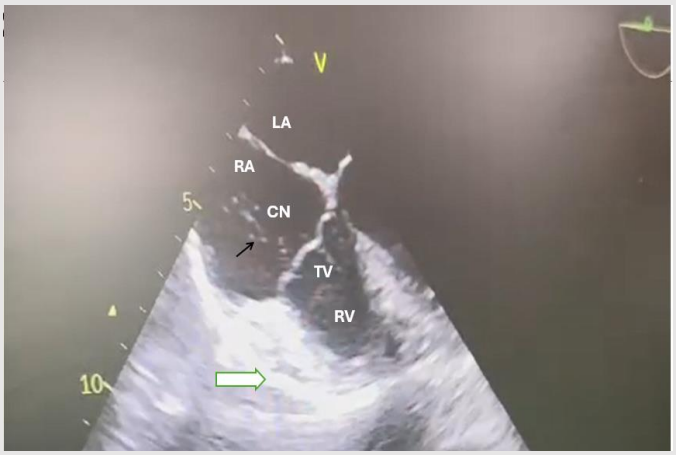

Figure 3: M-E Modified 4- C view of TEE. Shows multiple hyperechogenic strands of CN in the RA atrium (Black Arrow) and prolapsing into the tricuspid valve orifice. In addition, RV is hypertrophic (Thick white arrow), but TV appears morphologically normal, but some CN strands are seen protruding through the TV and resulting in the moderate TR in a normal TV
M-E -Modified 4- C view of TEE- mid oesophageal modified four chamber view, CN- Chiari Network, RA- right atrium, TV – tricuspid valve, TR- tricuspid regurgitation, RVH- right ventricular hypertrophy

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Video. 1. Color -Doppler Examination of the ME- modified Bicaval View shows TR and CN strands in RA. ME- mid -esophageal, TR – tricuspid regurgitation, CN- chiari Network.

Discussion:

Chiari Network is a rare cardiac anomaly in the right atrium, first time described by Hans Chiari, an Austrian pathologist in 1897, as a network or 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.[1] Basically, these are uncommon anatomical variants present as filamentous, weblike structure of the membrane of the fine fenestrated eustachian valve remnants persist in the RA, or the incomplete involution of the fetal sinus venosus valves results in redundant Chiari network.[2,3,4,5]

Its Prevalence in the general population varies from 2%-10% of randomly selected, asymptomatic patients.[6] Frequently, the Chiari network produces the diagnostic confusion on echocardiography as it depicts a mobile, net-like structures in right atrium near the opening of inferior vena cava and coronary sinus.[7] Sometimes the presence of a Chiari network is associated with tricuspid regurgitation, infectious endocarditis, migraine, embolic transient ischemic attack and/or stroke, in the presence of a patent foramen ovale.[7,8,9] Sometimes, CN might facilitate the

genesis of accessory pathway of atrioventricular conduction and causes Wolf–Parkinson–White syndrome.[7] Ali H et al have reported that CN and other RA remnants may cause entrapment of various devices or catheters during percutaneous cardiac procedures requiring right heart access.[10] However, this is usually of no clinical significance and often diagnosed incidentally when echocardiography is utilized to investigate the other cardiac defects.[11] Indeed, JP Chang et al and Joyce JJ et al and other authors have reported a significant TR due to the CN protruding into the RV. [11,12,13] some authors have diagnosed CN attaching to the septal leaflet of the tricuspid valve resulting in the central TR due to persistent prolapse of the leaflet and required the excision of CN under cardiopulmonary bypass.[11] In the presented patient, the CN was also an incidental finding of TEE which was performed to rule out the cardiac structural defect for the RVH and J point elevation revealed on the 12 lead ECG during routine preoperative assessment for cholecystectomy. This J point elevation suggest an early repolarization (ER) and may be associated with "Brugada syndrome" where right bundle branch block (RBBB) appears without an S wave, suggesting that in these cases the RBBB is apparent and that the R' represents an accentuation of the J wave. Some authors have reported ventricular fibrillation in patients with prominent J wave and ST-segment elevation in inferior leads without structural heart diseases and postulated that idiopathic ventricular fibrillation with ER pattern in inferior leads may represent a variant of the Brugada syndrome.[14,15]Therefore, it was decided to perform perioperative TEE in this patient for detection of structural cardiac defects and hemodynamic deterioration, and to guide the fluid and drug therapy. However, TEE revealed the CN that was protruding in the RV and causing moderate TR and RVH. The strands of CN were protruding through the tricuspid valve in diastole and affecting the coaptation during the systole resulting in TR, though morphologically tricuspid leaflets and annulus appear to be normal. [Figure 4] CN are hyperechoic membranous structures located within the RA, usually originates in the most posterior part of the (RA) and extends to a variable distance medially and inferiorly. However, the extent of Chiari network prolapse gradually decreases and many related abnormalities resolve during growth, but in the described patient the network prolapse persisted resulting in moderate TR even at the age of 22 years.

Conclusion:

Chiari Network usually considered as non-significant anomaly, and incidentally detected during the routine TEE evaluation in the patients undergoing various adult and paediatric cardiac surgical procedures. This single case report suggest that the possibility of Chiari network must be taken in the consideration for the patients presenting with ECG changes of RVH and RBBB. The caretaker must be aware of the possibility that Chiari Network may persist in the adult patients without regressing spontaneously. Rarely it may even protrude in the RV during RA contraction(diastole) and may cause TR even in morphologically normal leaflets and non-dilated tricuspid annulus. Therefore, Patients with ECG findings of RVH and RBBB should undergo regular echocardiography evaluation to rule out the structural cardiac defects including Chiari Network induced TR.

Funding: None.

Authorship: All authors have contributed equally to the design of the intellectual content of the case report and have approved the manuscript.

Conflict of interests:

The authors have no conflict of interest to declare.

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