Chiari Network - A diagnostic dilemma

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Abstract:
A male child of 4 years age, presented as a case of pyrexia of unknown origin, as a part of fever work up, on echocardiography was found to have hyperechoic flail mass inside right atrium attached to interatrial septum. Further investigations ruled out infective endocarditis and the mass was Chiari network a normal variant which poses a diagnostic dilemma in relevant clinical situation.

Keyword:
Pyrexia Of unknown Origin, hyperechoic mass in right atrium, Chiari network, infective endocarditis

Introduction:
Chiari network first described by Hans Chiari in 1897, a congenital remnant, seen in otherwise normal heart (1). It is present in 1.5% to 3% of normal population. This congenital remnant could be confused with valve disruption, vegetation or other mass lesion, particularly when associated with a suggestive clinical situation. The purpose of presenting this case is to emphasize Chiari network also should be considered in any echogenic mass in right atrium.

Case Report
A male child of 4 years age 2nd born of III degree consanguinous parents was brought with high grade continuous fever for 3 weeks. There was no history suggestive of any focus of infection. On examination, child was febrile and toxic with no localising signs. Investigations showed blood counts within normal limits (total count 7100 cells/cu mm, differential count P 43% L 52% E 7%, Hb 10.4g %, platelet 2 lakhs/cu mm), peripheral smear – moderately hypochromic anisopoikilocytosis & platelet in good clumps. Malarial parasite and microfilaria -negative, Urine albumin – Nil, 2 to 5 pus cells/ HPF, Culture – No growth, Blood culture – No growth, Chest Xray – Normal, USG abdomen showed hepatosplenomegaly. As a part of fever work up Echocardiogram was done which showed hyperechoic thread like structure seen in right atrial cavity measuring 10mm attached to interatrial septum, intracardiac anatomy being normal, no pericardial effusion. A differential diagnosis of vegetation, Chiari network was thought of. In order to rule out infective endocarditis antibiotics were
stopped for 48 hours and 3 blood cultures from 3 different sites were taken over a period of 24 hours. All three cultures were negative. Further investigations showed Widal negative, Mantoux negative, sputum for AFB negative, MSAT ++ confirmed by MAT. In view of leptospirosis, child was started on Inj.Crystalline penicillin and child showed dramatic response. The echogenic mass was chiari network which is a congenital remnant.

Discussion
Chiari network results from persistence of the valves of the embryonic systemic venous sinus. The dividing partition is placed between the systemic venous sinus and the distal part of the right atrium, made up of the vestibule and appendage. The embryonic valvar structures normally regress in late fetal life and early childhood. They persist as the Eustachian and Thebesian valves, the valves of the inferior caval vein and coronary sinus, respectively. These valves can retain their fetal proportions in abnormal conditions and then divide the right atrium. The dividing partition can itself be fenestrated so as not to produce major obstruction to the flow of blood. This is termed a Chiari network, and may not produce problems (2).
This structure can present as a highly mobile, highly reflectant echo target that can be seen in several locations in the right atrium. This congenital remnant, which is found pathologically in 2-3% of normal hearts, could be confused with valve disruption, vegetation or other mass lesion, particularly when associated with a suggestive clinical situation (3).

There have been reports of Chiari network in the right atrium being resected through a right atriotomy during aortic valve replacement for severe aortic stenosis (4).

Chiari networks have a well-documented association with a PFO. Although uncommon, Chiari networks can serve as a site for the formation of thrombus. This may be primary thrombus formation in the network or capture of venous thromboembolic matter. Right atrial thrombus may be asymptomatic, result in variable degrees of pulmonary emboli, or in association with a PFO may become a source of cerebral and peripheral arterial emboli (1).

Cardiac arrhythmias in association with Chiari networks also have been reported with termination of the arrhythmias being accomplished through surgical excision. There are also reports of Chiari networks acting as a physical barrier that interferes with the introduction of right-sided catheters and pacemakers (1). In our case of PUO Chiari network led to the confusion of infective endocarditis. Chiari networks are often picked up incidentally with echocardiography as an asymptomatic right atrial mass. Echocardiography, in particular transesophageal echocardiography, is both a reliable and noninvasive means of identifying Chiari networks. It should be considered in the differential diagnosis of endocarditis.

Reference:
2 Anderson RH, Baker EJ, Penny D, Redington AN, Rigby ML, Wernovsky G et al. Pediatric Cardiology, 3rd ed, Churchill livingstone Elseiver limited, 2010; Chapter 26, Division of Atrial chambers; p 550


Chiari Network - Echo