Chiari network: suboptimal imaging leading to diagnostic uncertainty and an adverse outcome

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Advances in cardiac diagnostic imaging have allowed an increased detection of both clinically relevant and incidental findings. Incidental findings may require individualized decisions which carry associated risks. We present a case in which an echocardiographic finding in an immunosuppressed patient changed the management and led to an adverse event.

A 48-year-old female with Crohn’s disease, on chronic adalimumab therapy, presented with a three-month history of generalized weakness, unintentional weight loss, night sweats, fevers, and chills. There was no history of recent central lines or intravenous drug use. She was admitted to the hospital with worsening hypoxemia and tachycardia. Chest computed tomography imaging and a transthoracic echocardiogram (TTE) were performed. The former revealed a miliary pulmonary infiltrate, and the latter identified a mobile, serpiginous, 2.5 cm mass in the right atrium, possibly attached to the Eustachian valve (Figure 1). The initial impression was that the echocardiographic mass could represent a thrombus, vegetation, or a Chiari network. Chiari network was thought to be unlikely due to the unusual thickness of the mass, as well as the clinical context of infection in an immunocompromised host. A transesophageal echocardiogram (TEE) was deferred given the patient’s worsening respiratory status. Due to concerns for a right atrium thrombus, IV heparin was initiated. Subsequently, the patient developed a massive retroperitoneal bleed. After stabilization, a TEE was performed as her respiratory status had significantly improved. The TEE elucidated a prominent, long mobile mass arising from the Eustachian valve (Figure 2). After further characterization of the mass, it became clear that it was a Chiari network with unusually increased dimensions. Eventually, the patient was diagnosed with disseminated tuberculosis and showed satisfactory response to antimicrobial therapy.

Discussion

Chiari’s network is an embryological remnant from the right sinus venosus valve named after Hans Chiari, who described the presence of meshwork-like tissue in the right atrium of cadavers in 1897.1 During the early embryonic period, systemic venous return converges to the sinus venosus. This cavity is connected to the right atrium (RA) by the sinoatrial orifice, which is surrounded by right and left venous valves. The right valve extends from the orifices of the inferior vena cava (IVC) to the superior vena cava and coronary sinus, and it directs oxygenated blood from the IVC to the left heart via the foramen ovale. Chiari’s network likely originates from failure of complete resorption of this valve.2–4

This embryonic remnant is found in 1.5–2% of patients.5,6 Echocardiographically, it typically appears as a thin, threadlike, very mobile echogenic mass usually attached on one end to the Eustachian orifice and the Thesbian valves, with the other end variably found in different right atrial positions.3,5–7

A Chiari network can be confounded with other right atrium masses.2,4–6 When atypical in appearance or when found in complex cases, differentiating a Chiari network from right atrial thrombus, vegetation, or ruptured tricuspid chordae tendineae can be challenging.6,9–12 Moreover, distinction between different causes can be influenced by the use of TTE versus TEE. In a study using both methods, only 28% of the TEE-identified Chiari networks were detected during TTE.5

Conclusion

In a patient with a complex clinical scenario, the choice of imaging modality may be limited by the patient’s medical conditions. This can lead to inadvertent clinical decisions based on available but incomplete imaging information. Our case illustrates one potential pitfall that can occur after using a suboptimal imaging modality for a right atrial mass.

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REFERENCES


Mid-esophageal right ventricle inflow-outflow view

The same mobile structure (arrow) is seen in the right atrium with one end attached to the Eustachian valve.

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