Giant Right Atrium Dissecting Aneurysm Mimicking Ebstein’s Anomaly

A 59-year-old male was referred to our department with palpitations for 1 month. Physical examination revealed a systolic murmur grade 3/6 heard at the tricuspid area. The electrocardiogram suggested ectopic atrial tachycardia. Chest x-ray showed a cardiomegaly with a bulging right cardiac border (Figure 1A).

Figure 1. (A) Chest x-ray shows cardiomegaly with a bulging right cardiac border. (B) TTE reveals a giant RA and a large aneurysmal sac communicating with RA, and the tricuspid valve was normally located. (C) CDFI shows multiple bidirectional shunting between the aneurysmal sac and RA. (D and E) CTA shows a giant aneurysm with low-density intimal flap-like structures arising from the RA. (F) CMR shows the giant aneurysm protruding to and overlapping on the surface of the RV. (G) The intraoperative findings confirm a giant RA dissecting aneurysm with a paper-thin wall. (H and I) Postoperative histopathological examination shows that the atrial wall consists of fibrin with fractured elastic fiber and several residual degenerative myocardial tissues. CDFI, Color Doppler flow imaging; CMR, cardiac magnetic resonance; CTA, cardiac tomography angiography; LA, left atrium; LV, left ventricle; RA, right atrium; RAA, right atrium appendage; RV, right ventricle; TTE, transthoracic echocardiography.
Transthoracic echocardiography (TTE) revealed a giant right atrium (RA) and a large aneurysmal sac communicating with RA, and the tricuspid valve was normally located (Figure 1B). Color Doppler flow imaging showed multiple bidirectional shunting between the aneurysmal sac and RA (Figure 1C). Cardiac tomography angiography (CTA; Figures 1D and E) and cardiac magnetic resonance (CMR) imaging (Figure 1F) showed a giant aneurysm with low-density intimal flap-like structures arising from the RA, which deformedly protruded to and overlapped on the surface of the right ventricle. The RA dissecting aneurysm was suspected based on these characteristics of multimodality images. The patient underwent surgical resection of the dissecting aneurysm, reconstruction of the RA wall, tricuspid valve ring annuloplasty, and modified Maze procedure with radiofrequency under extracorporeal circulation. The intraoperative findings confirmed a giant RA dissecting aneurysm with a paper-thin wall (Figure 1G). Postoperative histopathological examination showed that the atrial wall consisted of fibrin with fractured elastic fiber and several residual degenerative myocardial tissues (Figure 1H and I). The postoperative course was uneventful and was discharged 7 days later.

Giant RA dissecting aneurysm is a rare abnormality, which caused pseudo-displacement of the tricuspid valve and may easily be misdiagnosed as Ebstein’s anomaly, and it can also be confused with various anomalies, such as pericardial effusion, pericardial cysts, and tumors. Diagnosis of a RA dissecting aneurysm greatly depends on imaging modalities. Both TTE and transoesophageal echocardiography are very effective in discovering the aneurysm, distinguishing it from other abnormalities, such as pericardial cysts, Ebstein’s anomaly, and cor triatriatum, and detecting intracardiac thrombosis or blood flow limitations. Computed tomography or CMR imaging may help with more details on the neighboring structures of the aneurysm, especially when echocardiographic images are suboptimal. Due to the potential risk, some patients may develop complications including cardiac arrhythmia, thromboembolic events, congestive heart failure, and aneurysm rupture. Therefore, it is essential for correct diagnosis and appropriate treatment. Even though the optimal treatment strategy is still controversial, conservative treatment is always an option for asymptomatic patients, the surgical aneurysmectomy is an effective approach for the reduction of these life-threatening complications. We report a rare case of a giant RA dissecting aneurysm successfully treated by surgical procedures. Our case highlights that multimodality imaging plays an important role in distinguishing from other mimic anomalies and assessing this dissecting aneurysm location, size, and the relationship of surrounding adjacent structures. It can provide important clinical value for following surgical treatment.

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REFERENCES