An 11-year-old boy was referred to our hospital for near syncope. He was generally healthy before. On physical examination, his consciousness was clear, and there was no focal neurological deficit. The heart beat was irregular. ECG showed atrial flutter with variable ventricular response (Figure 1A). Cardiomegaly was noted on chest x-ray, with cardiac apex pointing toward the left side (Figure 1B). However, the transthoracic echocardiography showed that the location of presumed cardiac apex on chest x-ray was actually occupied by an extremely dilated left atrium (LA). The cardiac apex was pointed toward the right side of the chest. Mild to moderate mitral valve regurgitation was found, but the

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morphology and opening of both mitral leaflets appeared normal. Computed tomography confirmed the diagnosis of a giant LA aneurysm with rightward shift of the cardiac apex (Figure 1C and 1D). The LA appendage was not dilated (Figure 1D). Because atrial flutter was refractory to pharmacological conversion by intravenous amiodarone infusion, direct current cardioversion was performed and sinus rhythm was restored.

To prevent the formation of left-sided embolism and the recurrence of atrial flutter, aneurysmectomy was performed. A mitral ring was implanted at the same time. Both ventricles were deviated to the right side (Figure 2A). After aneurysmectomy, the heart regained its usual position immediately (Figure 2B). Grossly, the resected specimen appeared like a paper-thin strip of fibrous tissue (Figure 3A). Histological findings demonstrated focal thinning and irregular derangement of the myocardium with increased interstitial fibrosis (Figure 3B and 3C). Postoperative course was uneventful, and he was discharged 2 weeks after operation. The patient remained in normal sinus rhythm without medication for 4 months postoperatively. Follow-up chest x-ray showed normal cardiac silhouette (Figure 4A), and cardiac computed tomography demonstrated a usual-sized LA with normal position of the cardiac apex (Figure 4B and 4C).

Congenital LA aneurysms are extremely rare.1–4 In previously reported cases, most congenital LA aneurysms were extrapericardial and related to pericardial defects.1–3 In contrast, the intraoperative and histological findings of our present case demonstrated that the overlying pericardium of the LA aneurysm was intact. However, focal thinning of the aneurysm wall with irregular derangement of the myocardium was noted. Therefore, the formation of the intrapericardial LA aneurysm in this patient would be attributed to the congenital weakness of the atrial wall, resulting from intrinsic anomaly in the development of LA muscular layer combined with enhanced interstitial fibrosis.

Another unique finding of this case is that such a huge LA aneurysm significantly distorted the spatial distribution of the heart within the chest cavity. Therefore, the initial clinical suspicion was dextrocardia. Interestingly, the cardiac apex shifted to left side immediately after aneurysmectomy. To our knowledge, this striking feature has never been reported previously in the literature.

Figure 2. Intraoperative findings. A, A giant left atrial (LA) aneurysm occupied most of the space over left chest cavity, displacing both ventricles to the right side. B, After aneurysmectomy, the heart regained its usual position. LV indicates left ventricle; RA, right atrium; RV, right ventricle.

Figure 3. Histopathologic findings of the resected specimen. A, Macroscopic examination revealed a paper-thin, semitransparent membrane with cordlike structures on its endocardial surface. B, Hematoxylin and eosin staining showed focal thinning and irregular derangement of the myocardium (arrow heads). C, Masson trichrome staining demonstrated increased interstitial fibrosis (blue) with thinning myocardium (red).
Disclosures
None.

References


Figure 4. Postoperative image findings. A, Chest x-ray revealed normal heart size with normal cardiac silhouette. B and C, Chest computed tomography with 3-dimensional reconstruction showed marked reduction in left atrial (LA) size with resumed cardiac apex toward the left side (arrowheads). L indicates left; LV, left ventricle; R, right; and RV, right ventricle.
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