

Persistent Atrial Fibrillation Related to a Congenital Pericardial Defect and Left Atrial Appendage Herniation

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Congenital pericardial defects (CPDs) are infrequent anomalies that are usually asymptomatic and are discovered incidentally during unrelated interventions. Here we report the case of a CPD with herniation of an enlarged left atrial appendage identified during total thoracoscopic ablation (TTA) for persistent atrial fibrillation (AF). The persistent AF was successfully treated with a hybrid procedure, in which TTA was followed by an electrophysiological study.

Key words: 1. Pericardium
2. Atrial fibrillation
3. Video-assisted thoracic surgery
4. Herniation
5. Atrial appendage

CASE REPORT

Congenital pericardial defects (CPDs) are uncommon anomalies that are usually discovered unexpectedly by a cardiologist, radiologist, or surgeon. Occasionally, these defects may cause symptoms such as chest pain, dyspnea, arrhythmias, syncope, and even sudden death. Arrhythmias, including atrial fibrillation (AF), are uncommonly found in cases of CPD [1]. Herein, we report the case of a patient with persistent AF associated with a CPD and herniation of an enlarged left atrial (LA) appendage. The patient was treated successfully with a hybrid procedure, in which total thoracoscopic ablation (TTA) was followed by an electrophysiological (EP) study.

A 64-year-old male patient visited Samsung Medical Center complaining of palpitations. He had been treated for AF for two years. His medical history was otherwise unremarkable, except for a history of hypothyroidism diagnosed eight years

previously. A hybrid procedure involving TTA followed by an EP study was planned to treat his persistent AF. The pre-operative laboratory tests showed no abnormal findings. Preoperative computed tomography (CT) and echocardiography showed no evidence of thrombi or vegetation with LA enlargement. The size and volume index of the LA were 55 mm and 69.4 mL/m², respectively. TTA was performed under general anesthesia with double-lumen intubation. On the right side, a larger than normal atrium was identified (Fig. 1A). We performed ablation using methods described previously [2]. On the left side, a large pericardial defect was found, through which an enlarged LA appendage was herniated (Fig. 1B). Although handling the lighted dissector (AtriCure Lumitip Dissector; AtriCure Inc., Cincinnati, OH, USA) was difficult due to the large mobile LA appendage, pulmonary vein isolation and additional superior and inferior line ablation were successfully completed, and the LA appendage was

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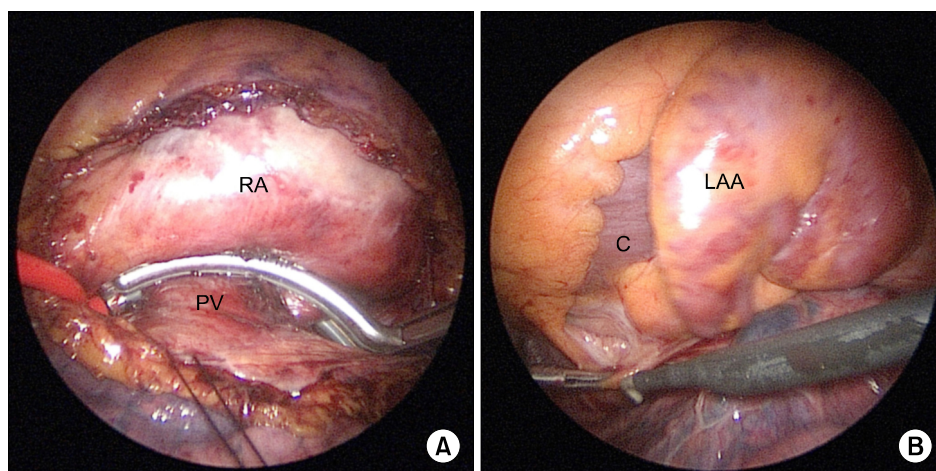


Fig. 1. Operative views of thoracoscopic ablation. (A) Right-side thoracoscopic view of the PV isolation. An enlarged atrial chamber is observed. (B) Left-side thoracoscopic view showing a congenital pericardial defect and a large LAA herniated through the defect. PV, pulmonary vein; RA, right atrium; C, congenital pericardial defect; LAA, left atrial appendage.

resected with an endoscopic stapler. An episode of AF occurred on the fourth postoperative day, and the patient was converted to sinus conversion through cardioversion. Otherwise, the patient's postoperative recovery was uneventful. A follow-up EP study on the eleventh postoperative day revealed successful pulmonary vein isolation, and cavotricuspid isthmus ablation was performed. The patient was discharged on the twelfth postoperative day.

DISCUSSION

CPDs are rare anomalies derived from improper development of the pleuropericardial membrane [3]. They range in extent from partial defects to complete defects of the pericardium. Left-side defects are most common, accounting for approximately 70% of cases, followed by right-side defects (17%) and complete defects (9%) [4].

Although patients are usually asymptomatic and most CPDs are discovered incidentally, patients with partial defects may present with acute symptoms such as chest pain, dyspnea, arrhythmia, syncope, and even sudden death due to strangulation of the herniated cardiac chamber [5].

CPDs are difficult to diagnose. Echocardiography and CT are the most common methods used to evaluate cardiac and thoracic abnormalities; however, pericardial defects may go unrecognized for years, as in this case [1]. Magnetic resonance imaging (MRI) has been reported to be superior to echocardiography [6], but cardiac MRI is still only applicable to a limited set of lesions.

The relationship between AF and CPD is not well understood. Herniated LA appendages have been suggested to be a cause of paroxysmal AF [1]. Furthermore, enlargement of the LA and LA appendages due to herniation, as in our patient, may contribute to the development of persistent AF, and may even be its cause [7].

Several methods have been proposed for the treatment of CPD, including LA appendectomy, extension of the pericardium to reduce incarceration, primary closure, and patch closure using synthetic materials [1,8,9]. However, no consensus currently exists regarding therapeutic options for CPD due to the small number of cases. Although a previous case of paroxysmal AF was resolved via resection of the LA appendage instead of antiarrhythmic interventions [1], in cases of persistent AF with an enlarged chamber, as in our patient, antiarrhythmic management techniques such as TTA are probably necessary.

The Cox-Maze III procedure is considered the gold standard treatment for AF. However, it involves complex techniques and invasive procedures, requiring median sternotomy and cardiopulmonary bypass [10-12]. Accordingly, some researchers have recently suggested that minimally invasive thoracoscopic ablation be used, and hybrid TTA has shown excellent results with low morbidity rates [13-15]. In our patient, the hybrid TTA procedure was initially planned for his persistent AF, and the CPD was an incidental finding. Although the visual field was disturbed by the enlarged LA appendage, the entire ablation procedure was successfully performed and the LA appendage was resected safely.

In conclusion, CPD is a possible incidental finding when performing cardiac operations, including TTA. Enlarged LA and LA appendages due to herniation through the defect may cause persistent AF, and antiarrhythmic treatment may be necessary for such patients. TTA can be successfully performed in patients with persistent AF associated with a CPD and herniation of the enlarged LA and/or an LA appendage.

CONFLICT OF INTEREST

No potential conflict of interest relevant to this article was reported.

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