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 preoperative 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...
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 antarrhythmic 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.

**REFERENCES**

1. Misthos P, Neofotistos K, Drosos P, Kokotsakis J, Lioulias A. Paroxysmal atrial fibrillation due to left atrial appendage herniation and review of the literature. Int J Cardiol 2009; 133:e122-4.

2. Lee HM, Chung SR, Jeong DS. Initial experience with total thoracoscopic ablation. Korean J Thorac Cardiovasc Surg 2014; 47:1-5.

3. Ellis K, Leeds NE, Himmelstein A. Congenital deficiencies in the parietal pericardium: a review with 2 new cases including successful diagnosis by plain roentgenography. Am J Roentgenol Radium Ther Nucl Med 1959;82:125-37.

4. Centola M, Longo M, De Marco F, Cremonesi G, Marconi M, Danzi GB. Does echocardiography play a role in the clinical diagnosis of congenital absence of pericardium?: a case presentation and a systematic review. J Cardiovasc Med (Hagerstown) 2009;10:687-92.

5. Robin E, Ganguy SN, Fowler MS. Strangulation of the left atrial appendage through a congenital partial pericardial defect. Chest 1975;67:354-5.

6. Cuccuini M, Lisi F, Consoli A, et al. Congenital defects of pericardium: case reports and review of literature. Ital J Anat Embryol 2013;118:136-50.

7. Parkash R, Green MS, Kerr CR, et al. The association of left atrial size and occurrence of atrial fibrillation: a prospective cohort study from the Canadian Registry of Atrial Fibrillation. Am Heart J 2004;148:649-54.

8. Juarez AL, Akerstrom F, Alguacil AM, Gonzalez BS. Congenital partial absence of the pericardium in a young man with atypical chest pain. World J Cardiol 2013;5:12-4.

9. Hoornije JC, Mooyaart EL, Meuzelaar KJ. Left atrial herniation through a partial pericardial defect: a rare cause of syncope. Pacing Clin Electrophysiol 1989;12:1841-5.

10. European Heart Rhythm Association (EHRA); European Cardiac Arrhythmia Society (ECAS); American College of Cardiology (ACC), et al. HRS/EHRA/ECAS expert Consensus Statement on catheter and surgical ablation of atrial fibrillation: recommendations for personnel, policy, procedures and follow-up: a report of the Heart Rhythm Society (HRS) Task Force on catheter and surgical ablation of atrial fibrillation. Heart Rhythm 2007;4:816-61.

11. Prasad SM, Maniar HS, Camillo CJ, et al. The Cox maze III procedure for atrial fibrillation: long-term efficacy in patients undergoing lone versus concomitant procedures. J Thorac Cardiovasc Surg 2003;126:1822-8.

12. Raanani E, Albage A, David TE, Yau TM, Armstrong S. The efficacy of the Cox/maze procedure combined with mitral valve surgery: a matched control study. Eur J Cardiothorac Surg 2001;19:438-42.

13. Bisleri G, Curnis A, Bottio T, Mascioli G, Muneretto C. The need of a hybrid approach for the treatment of atrial fibrillation. Heart Surg Forum 2005;8:E26-30.

14. Pison L, La Meir M, van Opstal J, Blauw Y, Maessen J, Crijns HJ. Hybrid thoracoscopic surgical and transvenous catheter ablation of atrial fibrillation. J Am Coll Cardiol 2012; 60:54-61.

15. Muneretto C, Bisleri G, Bonenti L, Curnis A. Durable staged hybrid ablation with thoracoscopic and percutaneous approach for treatment of long-standing atrial fibrillation: a 30-month assessment with continuous monitoring. J Thorac Cardiovasc Surg 2012;144:1460-5.