Cor triatriatum in adulthood with mitral valve regurgitation and atrial fibrillation

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Abstract

Cor triatriatum is a rare congenital heart disease. A 57-years-old woman had cor triatriatum with severe mitral valve regurgitation (MR) and atrial fibrillation (AF). We perfomed mitral valve repair, left atrial appendage resection, and maze procedure by resection of the anomalous septum in the left atrium. At result, MR was controllable and AF disappeared after the operation. Although there is no established maze procedure with cor triatriatum, removing the septum was effective to complete it.

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Cor triatriatum in adulthood with mitral valve regurgitation and atrial fibrillation

Authors:

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Abstract

Cor triatriatum is a rare congenital heart disease. A 57-years-old woman had cor triatriatum with severe mitral valve regurgitation (MR) and atrial fibrillation (AF). We perfomed mitral valve repair, left atrial appendage resection, and maze procedure by resection of the anomalous septum in the left atrium. At result, MR was controllable and AF disappeared after the operation. Although there is no established maze procedure with cor triatriatum, removing the septum was effective to complete it.

A 57-year-old woman was transported to our hospital for ventricular fibrillation after defibrillation by automated external defibrillator. Emergency coronary angiography revealed no significant stenosis. Transthoracic

echocardiography showed severe MR and we initiated treatment for heart failure caused by MR with cerebral hypothermia. The treatment was successful, and the heart failure seemed well controlled by medical therapy. However, severe MR remained, and AF developed during the course of treatment. We performed mitral valve repair with the maze procedure. Preoperative echocardiography revealed mitral valve P2 prolapse; additionally, an anomalous septum was found in the left atrium, which proved to be a cor triatriatum (Figure 1). Enhanced computed tomography showed that the accessory chamber and the left atrium communicated via a 7-mm hole, and the four pulmonary veins entered the heart through the accessory chamber (Figure 2). Through a median sternotomy, we performed mitral valve repair, left atrial appendage resection, and maze surgery. Approaching the accessory chamber by right lateral left atriotomy, we excised as much of the anomalous septum as possible (Figure 3). The maze procedure was completed by radiofrequency ablation across the remaining part of the septum (Figure 4). For MR, we performed annuloplasty using a ring and artificial tendon reconstruction. The postoperative course was uneventful; MR was controllable and AF disappeared.

Cor triatriatum is a rare congenital heart disease. It is usually diagnosed during childhood; a new diagnosis in adults is rare, and cor triatriatum alone may not be an indication for surgery[1]. In this case, because of severe MR and AF, we performed the operation. There is no established maze procedure for cor triatriatum [2]. We completed the surgery by removing the septum; consequently, atrial fibrillation disappeared.

Author contributions

S.I., T.F., K.Y. and M.M. designed and performed the experiments, analysed data and interpreted it. S.I. and K.Y. Drafted article. S.I., T.F., K.Y. and M.M. revised it critically. S.I., T.F., K.Y. and M.M. approved of the article, collected data and supported technical and logistical.

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Figure 1. Transthoracic echocardiography, showed a septum (arrow) in the left atrium. LA: left atrium, LV: left ventricular.

Figure 2. Preoperative enhanced computed tomography. A, The accessory chamber was separated from the left atrium by the anomalous septum. B, The accessory chamber and the left atrium communicated via a 7-mm hole (yellow arrow). C, All four pulmonary veins entered the heart through the accessory chamber.

Figure 3. Intraoperative pictures taken from the surgeon's perspective. A, The septum separating the left atrium and the hole (arrow) was found. B, After excising the septum, the mitral valve was visually recognized (asterisk: posterior leaflet of the mitral valve).

Figure 4. The left atrial maze procedure schema. The anomalous septum was excised, and an ablation of MV isthmus line was performed across the remained septum after resection. LA: left atrium, LPA: left pulmonary artery, RPA: right pulomonary artery, MV: mitral valve.