Membranous Septal Aneurysm: An Unusual Case for Sub-Pulmonary Obstruction in cTGA

Membranöz Septal Anevrizma: cTGA'da Subpulmoner Obstrüksiyonun Nadir Bir Nedeni

cTGA Membranöz Septal Anevrizma Birlikteliği / cTGA with Membranous Septal Aneurysm

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Özet

İnterventriküler septumun membranöz bölümünün anevrizması nadiren tanı alan bir konjenital kardiyak malformasyondur. Perimembranöz ventriküler septal defektler en sık defekt tipini oluştururlar ve morfolojik olarak membranöz septumda görülürler. Triküspit septal liflet komşuluğundaki membranöz septal anevrizmaların cerrahi eksplorasyonu, onarımı triküspit kapak fonksiyonu ve iletim dokusuna yakınlığı nedeniyle önem arz eder. Bu çalışmada konjenital olarak düzeltilmiş büyük arter transpozisyonlu hastada subpulmoner obstriksiyona sebep olan membranöz septal anevrizmanın başarılı cerrahi tedavisini sunduk.

Anahtar Kelimeler

cTGA; Membranöz Septal Anevrizma; Pulmoner Stenoz

Abstract

Aneurysm of the membranous portion of the interventricular septum is an uncommon congenital cardiac malformation that is rarely diagnosed during life. Perimembranous ventricular septal defects are the most common type of the ventricular septal defects and originates from morphologically membranous interventricular septum. Surgical exposure and accurate closure of a ventricular septal defect with a membranous septal aneurysm beneath the tricuspid septal leaflet carries a risk of tricuspid valve insufficiency and conduction disturbances. The current study presents a case with membranous septal aneursym with congenitally corrected transposition of the great arteries and sub-pulmonary obstruction, which was surgically corrected.

Keywords

cTGA; Membranous Septal Aneursym; Pulmonary Stenosis

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Introduction

Aneurysm of the membranous portion of the interventricular septum is an uncommon congenital cardiac malformation, rarely diagnosed during life. Perimembranous ventricular septal defects are the most common type of the ventricular septal defects and originates from morphologically membranous interventricular septum. It has been known for several years that this variety of ventricular septal defect (VSD) is frequently associated with septal aneurysm formation. A membranous septal aneursym (MSA) has been defined as an abnormality having the angiographic appearance of a pouch-like structure with distinct margins protruding around the edges of a VSD and bulging into the right ventricle during systole. Surgical exposure and accurate closure of a VSD with a membranous septal aneurysm beneath the tricuspid septal leaflet carries a risk of tricuspid valve insufficiency and conduction disturbances. The current study presents a case membranous septal aneursym with congenitally corrected transposition of great arteries (cTGA) and sub-pulmonary obstruction, which was surgically corrected.

Case Report

An 11-year-old girl was referred to our clinic because of heart murmur and dyspnea on exertion. She had normal growth and development without cyanosis and congestive heart failure. Her medical history revealed frequent respiratory infections, but there was no history of bacterial endocarditis, rheumatic fever, or cardiac arrhythmias. Prior to admission, she had noticed progressive shortness of breath on exertion.

The physical examination revealed a grade 4/6 ejection murmur along the left fourth intercostal space and there was also a 3/6 pansystolic ejection murmur at the apex of the heart radiating to the sternum. Blood pressure was 135/90 mmHg and heart rate was 90 beats/minute. The electrocardiogram showed sinus rhythm and normal axis. The chest x-ray revealed moderate cardiomegaly. Transthoracic two-dimensional echocardiography revealed patent foramen ovale (PFO), atrio-ventricular and ventriculo-arterial discordance. Aorta was positioned on the left and anterior side of the pulmonary artery and there was a right aortic arch. Tricuspid and mitral valve coaptations were normal. Color flow and doppler examinations were showed a sub-pulmonary obstruction with 80 mmHg gradient at the valve site. There was no VSD in the doppler flow examination. Right and left heart catheterizations were performed. Catheterization confirmed the congenitally corrected transposition of the great arteries (cTGA) diagnosis. Morphologically left ventricle systolic pressure was 140 mmHg, and pulmonary artery systolic pressure was 30 mmHg with a 110 mmHg sub-pulmonary gradient. Morphologically right ventricle angiogram demonstrated an aneursym of the septum bulging into the sub-pulmonary region (Figure 1). Surgery was planned to relieve the sub-pulmonary obstruction. Median sternotomy and aortic-bicaval cannulation was performed. Cardiopulmonary bypass was instituted with moderate hypothermia, and antegrade crystalloid potassium cardioplegic solution was used. After cross clamping, the pulmonary artery was transversely incised. At the subvalvular area, fibrotic tissue was excised (Figure 2). After the excision of the MSA, a perimembranous ventricular septal defect was observed (Figure 3). The VSD was closed via pulmonary arteriotomy with double velour Dacron patch using the continuous technique (Figure 4). The patient was weaned from cardiopulmonary the bypass with normal sinus rhythm. After an uneventful recovery period, the patient was discharged on the postoperative fifth day.

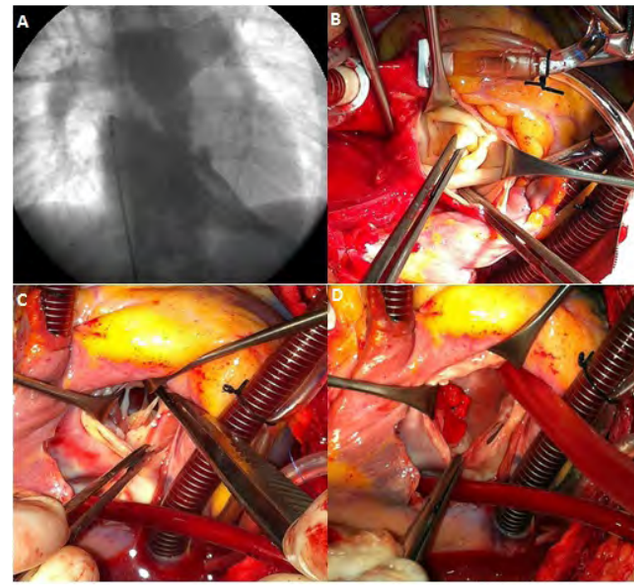


Figure 1. A. An angiographic image of membranous septal aneurysm, B. An intraoperative view of subpulmonary membraneus septal aneurysm, C. An intraoperative, view of ventricular septal defect after removing the membranues septal aneurysm, D. An intraoperative view of repaired ventricular septal defect

Discussion

MSA is a rare lesion that was found in 0.3 % of all congenital heart defects and was found in 19.1% of the patients who were examined for VSD [1].

Although most cases do not manifest themselves symptomatically, MSA can be the cause of systemic embolies, endocarditis, cardiac arrhythmias, left or right ventricular outflow tract obstruction, and right to left shunts secondary to ruptures.

MSA related sub-pulmonary obstruction was related in part to the intraventricular anatomy in these patients. With normal anatomy, it is unlikely that a MSA causes severe obstruction in the subpulmonary region. In contrast, in patients with cTGA, the pulmonary valve is usually in continuity with the right sided atrioventricular valve. The perimembranous ventricular septal defect is adjacent and immediately inferior to the pulmonary valve and there is no interposed crista supraventricularis. When MSA occurs in this anomaly, it is likely to cause a significant sub-pulmonary obstruction [2].

Although this defect seems small because the aneursym limits the shunt between the ventricles, the actual VSD tends to be larger than it was estimated. It is critical to open the aneursym to expose the true margin of the VSD for proper repair. In the literature, radial incision of the MSA or circular detachment of the tricuspid valve techniques were applied for this anomaly. But in this case it was prefered to excite the MSA by pulmonary arteriotomy and close the VSD via this arteriotomy [3,4].

A double velour Dacron patch was chosen to close the VSD because of the margins of the VSD were far enough from the pulmonary valve and there was a clear suture line in the subpulmonary area. However, in some instances there may not be a suture line in the subpulmonary area and while closing the VSD,

the pulmonary valve leaflets can be utilized to place the VSD closure stiches. In these cases, the patches that cause fibrotic reactions must be avoided for pulmonary valve functions. Additionally, if a fibrous suture line remains in place when resecting the aneursym, closing the VSD becomes more easy and safe because the atrioventricular node lies along the antero-superior margin, which is an unusual and vulnerable place.

Aneursym formation may be important because it reduces the functional size of an associated membranous VSD; on the other hand the aneursym itself is associated with some severe complications. Additional surgical intervention was not observed in other series concerning the surgical treatment of MSA [5]. Therefore, we can recommend that the MSA should be operated on during childhood period in order to prevent further enlargement and subsequent complications.

Competing interests

The authors declare that they have no competing interests.

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