be much higher. Death due to aneurysm rupture has been reported to occur even less than 4 weeks after the initial operation [3]. If 1-stage repair is possible, it is preferable, especially in patients with severely dilated or ruptured aneurysms. We believe that a 2-stage operation would not have been the best option for rescue in our case.

Recently, endovascular repair has been reported to be an effective alternative to open repair for ruptured aortic aneurysms. However, enlargement of residual aneurysm after primary aortic endovascular treatment has been reported, especially in patients with MFS [5]; thus, the long-term results of endovascular repair are unclear. In addition, endovascular repair for rupture of both the aortic root and TAAA is not possible.

When performing replacements of distant aortic areas, preparation for spontaneous rupture is crucial due to the long duration of CPB. Arterial cannulation must be carried out through both upper and lower body arteries, and the surgeon must be prepared for emergent aortic cross-clamping in every region. Systemic cooling can be resumed, even after cross-clamping for control of unexpected bleeding. In aortic surgery that requires a prolonged operation time secure organ protection is the key in preventing malperfusion-related postoperative complications. As for perfusion pumps, some surgeons prefer only 1 additional arterial pump for SCP or visceral perfusion to simplify CPB. However, we insist that 2 additional pumps be available in those cases to provide an adequate amount of blood flow for each dissected vessel, and preparation for possible emergent simultaneous SCP and selective visceral perfusion is crucial.

In conclusion, the single-stage replacement of the aortic root, arch, and thoracoabdominal aorta can be performed safely. Preparation for unexpected hemorrhage and adequate organ perfusion are key for saving patients who need extensive and multiple aortic operations. To the best of our knowledge, this is the first successful case of the simultaneous repair of multiple aortic aneurysms in a MFS patient with ruptured remote aneurysms.

References


Calcified Amorphous Tumor of the Left Ventricular Outflow Tract

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We report a very rare case of a calcified amorphous tumor presenting atypically as a mobile left ventricular outflow tract mass in a 69-year-old female who was admitted for shortness of breath.


Calcified amorphous tumors (CAT) of the heart represent a rare non-neoplastic tumor, first described by Reynolds and colleagues in 1997 [1]. They are most often noted to occur in the setting of exuberant mitral annular calcification (MAC) and end-stage renal disease [2]. We report a case of CAT presenting atypically in the left ventricular outflow tract (LVOT).

A 69-year-old female with history of chronic kidney disease was admitted for epigastric pain and shortness of breath. She was in a volume overload state requiring initiation of hemodialysis and underwent cardiac evaluation. Transthoracic echocardiogram revealed a normal ejection fraction of 0.60. Heavy MAC, with trace mitral regurgitation was seen. An echo dense mobile mass was visualized in the LVOT. There was no hemodynamic evidence of LVOT obstruction on Doppler echocardiography. Transesophageal echocardiogram showed the mobile mass attached to the LVOT in the region of the anterior mitral valve annulus (Figs 1, 2). The patient was not febrile and laboratory parameters did not reveal any elevated inflammatory markers. There was no history of malignancy or rheumatologic disorder. Cardiac catheterization showed mild left anterior descending coronary artery disease and 90% right coronary artery stenosis. Due to of the mass’ mobility and potential for embolism, surgical removal with concomitant single-vessel coronary artery bypass grafting was advised. Surgery was performed through a median sternotomy on cardiopulmonary bypass. After aortic cross-clamping and grafting of the right coronary artery, the mass was approached though an oblique incision of the ascending aorta toward the noncoronary sinus. A 5-m 30-degree angle angioscope attached to a 1288 HD Camera (Styker, Kalamazoo, MI) was advanced into the LVOT. The endoscopic view provided clear visualization of the
pedunculated mass attached to the ventricular surface to the anterior mitral valve leaflet and facilitated protection of the aortic valve. The mass was sharply excised from the mitral valve (Fig 3) Pathologic examination of the 0.7 × 0.3 cm mass demonstrated tissue with excess fibrin and calcium (Fig 4), consistent with a diagnosis of CAT of the heart. The postoperative course was uneventful with the patient discharged on postoperative day 8.

Comment
Calcified amorphous tumors are rare non-neoplastic cardiac masses composed of amorphous calcified material. They are often reported to be associated with the mitral valve, particularly in end-stage renal disease patients with exuberant MAC [1–3]. These lesions are often mobile and are difficult to differentiate from other lesions such as fibroelastoma, myxoma, or marantic vegetation on noninvasive imaging [4]. Although the exact etiology of CAT remains unclear, it is postulated that abnormalities of calcium, phosphorus, and vitamin D3, especially in patients with hemodialysis, may be responsible for their proliferation [2].

Excision of the lesions is generally curative, although recurrence has been reported after surgical excision, especially in the setting of incomplete resection [5]. Histologically, CAT can be distinguished from other lesions as they are composed of calcified nodules in an

Fig 1. Transesophageal echocardiography appearance of calcified amorphous tumor (arrow).

Fig 2. Three-dimensional transesophageal echocardiographic appearance of calcified amorphous tumor (arrow).

Fig 3. Angioscopic visualization of calcified amorphous tumor.

Fig 4. Histologic appearance of the tumor demonstrating presence of fibrin and calcium (original magnification ×40).
amorphous background of fibrin with degeneration and focal inflammation [3].

Calciﬁed amorphous tumors can present with symptoms of obstruction or embolization [6, 7]. The presence of MAC was noted to double the risk of stroke or transient ischemic attack in the Framingham cohort [8]. Embolization of fronds of MAC associated CAT may contribute to such an observation, although such link has not been conclusively determined.

Our case illustrates an atypical location of this nonmalignant, rare and unusual intracardiac tumor. Such tumors have a potential for embolization leading to stroke or transient ischemic attack, and surgical removal is usually curative. Our case was facilitated by use of intraoperative angioscopy, which resulted in complete removal of the mass reducing the likelihood of potential cardioembolic events in this patient.

References

Modified Nikaidoh Procedure With Double-Root Translocation in a 1-Year-Old Boy
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A 1-year-old boy weighing 10.4 kg underwent successful biventricular repair for transposition of the great arteries, a ventricular septal defect, and a left ventricular outﬂow tract (LVOT) obstruction with moderate pulmonary stenosis of the bicuspid pulmonary valve (z score of −4.4 for the pulmonary valve) by means of a modiﬁed Nikaidoh procedure with double root translocation by use of a valve-spared pulmonary root. The postoperative echocardiogram showed no LVOT obstruction, no aortic valve regurgitation, and mild pulmonary stenosis and pulmonary valve regurgitation. No reintervention has been required during the 6-year follow-up, with annular growth of the pulmonary valve.


Rastelli and réparation à l’etage ventriculaire (REV) procedures are the most common surgical procedures for transposition of the great arteries (TGA), ventricular septal defects (VSD), and left ventricular outﬂow tract (LVOT) obstructions with or without more than mild pulmonary stenosis. However, more aggressive procedures, including aortic root translocation (eg, the original Nikaidoh procedure) and double root translocation (DRT), have recently become important treatments of choice [1, 2]. We report our experience with a modiﬁed Nikaidoh procedure with DRT using a valve-spared pulmonary root.

A boy 1 year and 8 months old, weighing 10.4 kg, was referred to our institution for surgical repair after undergoing a left modiﬁed Blalock-Taussig shunt at 2 months of age. The preoperative examinations revealed a borderline cardiac anomaly between the TGA and a double-outlet right ventricle, VSD with an inlet extension considered to require repair for a remote-type VSD, and an LVOT obstruction with a 65 mm Hg pressure gradient, involving a bicuspid and tethered pulmonary valve (diameter, 7.4 mm; z score, −4.4). The aortic valve also had a 7.4-mm diameter (z score, −4.6).

The patient underwent a modiﬁed Nikaidoh procedure with aortic root translocation with use of a pivotal rotation technique, pulmonary root translocation with a valve-spared pulmonary root, and an arterial switch procedure with the Lecompte maneuver in 2006.

After cardiac arrest was induced in the usual manner, the pulmonary valve was checked through a longitudinal incision on the main pulmonary artery. This examination conﬁrmed that a neoaortic valve would be diﬃcult to use because of the size and severely fused bicuspid condition. We performed a modiﬁed Nikaidoh procedure with pulmonary root translocation (Fig 1). The cardiopulmonary bypass time and aortic clamping time were 323 and 171 minutes, respectively. The patient had a smooth postoperative course and was extubated the next day. Cardiac catheterization 1 year later conﬁrmed a good LVOT without aortic insuﬃciency and an acceptable right ventricular outﬂow tract (Fig 2). Moreover, it revealed minor pulmonary regurgitation and 20 mm Hg pressure gradient at his bicuspid pulmonary valve.