



Transaortic accessory mitral valve excision in an adult patient: Case report

Erişkin bir hastada transaortik yolla aksesuar mitral kapak eksizyonu: Olgu sunumu

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ABSTRACT

Accessory mitral valve tissue is a rare congenital cardiac anomaly, and is usually incidentally detected in childhood. In this case, we present a 65-year-old man with aortic stenosis originating from an accessory mitral valve leaflet attached to the anterior mitral leaflet. Interestingly enough, the patient's accessory mitral valve remained undetected for years until he became symptomatic for degenerative aortic stenosis. Aortic valve replacement and excision of the accessory mitral valve attached to the anterior mitral leaflet was performed with a transaortic approach instead of atriotomy. It was also unusual to see accessory mitral valve tissue with aortic stenosis instead of other frequently associated congenital anomalies.

Keywords: Accessory mitral valve; aortic stenosis; left ventricular outflow tract obstruction.

Accessory mitral valve (AMV) tissue is a rare congenital cardiac anomaly and is usually diagnosed incidentally for complaints such as shortness of breath, fainting, chest pain, palpitation, and fatigue during childhood. In some cases, it may lead to left ventricular outflow tract (LVOT) obstruction. Accessory mitral valve tissue is frequently associated with ventricular septal defects, subaortic stenosis, large vessel anomalies and atrial septal defects.^[1] The incidence of AMV in adults is estimated to be 1/26,000.^[2] However, the prevalence of AMV and the age in which it is most frequently diagnosed are still debated.^[3]

Although the exact embryological mechanism of AMV formation is unclear, it may be caused by an abnormal or incomplete separation of the endocardial cushions.^[4] Surgery is usually performed because of severe LVOT obstruction or other cardiac reasons.^[1]

ÖZ

Aksesuar mitral kapak dokusu, nadir bir doğuştan kardiyak anomalidir ve genellikle çocukluk çağında rastlantısal olarak tespit edilmektedir. Bu yazıda anterior mitral kapağa tutunmuş aksesuar mitral kapak ile birlikte aort darlığı olan 65 yaşında bir erkek hasta sunuldu. Hastanın uzun yıllar asemptomatik kalması ve dejeneratif aort darlığı semptomatik hale gelene kadar aksesuar mitral kapağın tespit edilememiş olması ilginçtir. Aort kapak replasmanı ve mitral anterior yaprakçığa tutunmuş aksesuar mitral kapak eksizyonu atriyotomi yerine transaortik yaklaşımla uygulandı. Aksesuar mitral kapağın sık eşlik ettiği diğer anomaliler yerine aort darlığı ile birlikte görülmesi yine alışılmadık bir durumdur.

Anahtar sözcükler: Aksesuar mitral kapak; aort darlığı; sol ventrikül çıkış yolu tıkanıklığı.

CASE REPORT

A 65-year-old male patient presented to our clinic with complaints of exercise intolerance, palpitations, and syncope. He had a history of stenting of the left anterior descending coronary artery due to acute coronary syndrome seven years previously. At that time, no valvular pathology had been detected. Physical examination revealed a 3/6 systolic murmur over the aortic area. Other physical examination findings and laboratory tests were normal. The patient was on amlodipine and ramipril treatment for three years. Transthoracic echocardiography revealed 60% left ventricular ejection fraction. The ascending aorta was 3.3 cm and the LVOT was 2.2 cm in diameter. The aortic valve was tricuspid in nature, thick, and calcified with maximum 65 mmHg and mean 36 mmHg transvalvular gradients. Planimetry showed

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Figure 1. Transesophageal echocardiographic view of the accessory mitral valve.

that aortic valve area was 1.1 cm². First degree aortic regurgitation with minimal mitral regurgitation was detected.

There was also a mobile thread-like structure attached to the mitral anterior leaflet at the LVOT and its echogenicity was similar to the mitral valve (MV). The fixation area of this structure showed coherence with the cord or papillary muscle. No other congenital anomalies were detected. Preoperative coronary angiography confirmed patent left anterior descending stent. A surgery was planned for aortic stenosis (AS) and aberrant tissue over the MV. Perioperative transesophageal echocardiography (TEE) was performed under general anesthesia and showed a MV-like structure in the LVOT (Figure 1). An echogenic parachute-like structure was present over the anterior MV with dimensions of 35×18×8 mm at the ventricular window. This structure was more prominent at peak systole resulting in turbulence in the LVOT. A transverse aortotomy was performed 2 cm above the level of the aortic valve. Through the aortic root, AMV was seen in the superior of the ventricular aspect of the anterior MV, in the lower part of the left leaflet of the aortic valve, and in the annulus with two tendinous chord lesions (Figure 2). The AMV leaflets were explored through the aortotomy in the LVOT and excised without giving damage to other MV structures. A number 19 mechanical valve was replaced in the aortic position. Post-cardiopulmonary bypass TEE showed that the AMV disappeared and the mitral and prosthetic aortic valve functioned normally. The postoperative period was uneventful. The patient was discharged on fifth day with warfarin treatment. Postoperative TTE was normal.



Figure 2. Perioperative accessory mitral valve tissue image through the aortotomy incision.

DISCUSSION

Accessory mitral valve is often associated with congenital cardiac anomalies in the pediatric patient group. In adult patients, AMV may be a clinical condition arising from the remote side, or it may be diagnosed incidentally. The exact prevalence of this anomaly is unknown because several cases of AMV were incidentally diagnosed intraoperatively.^[2]

As seen in this case, congenital AMV can remain asymptomatic for many years. The literature reports symptoms including dyspnea, syncope, chest pain, palpitations and arrhythmias, cerebrovascular accidents, low cardiac output due to subaortic obstruction, and congestive heart failure.^[1,2]

In symptomatic patients and patients who were scheduled for cardiac surgery for different pathologies, accessory valve resection is absolutely recommended.^[3] For isolated AMV, the current approach is intervention in patients with significant LVOT gradient (mean gradient of ≥ 25 mmHg). Patients with LVOT mean gradient under 25 mmHg, are recommended to be followed-up at regular intervals.^[1,2]

Although TTE is sufficient for diagnosis, perioperative TEE is helpful for both confirmation of the diagnosis and evaluation of the MV functions after excision.^[5,6] Transesophageal echocardiography is also important in the evaluation of possible complications that may occur after excision and revision of cardiopulmonary bypass for repair and completion of the surgical procedure.

Despite limited experience in the fields of AMV, and frequency of surgical treatment, and methodology

in the literature, it is stated that AMV excision could be performed via right or left atriotomy.^[1] In this case, aortic valve replacement was planned with aortotomy. The AMV tissue was easily accessed through the aortotomy side. We were able to perform excision of the AMV without the need for atriotomy.

As a result, in patients with accessory mitral valve tissue undergoing aortic surgery, the accessory valve can be easily removed through aortotomy without opening another heart chamber; therefore, potential systemic side effects due to extension of extracorporeal circulation can be avoided.

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