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Asymptomatic and isolated accessory mitral valve tissue in adult population: three case reports and review of the literature

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Abstract. – Accessory mitral valve tissue is a rare congenital cardiac anomaly and commonly it may cause left ventricular outflow tract obstruction (LVOTO). This anomaly occurs as a part of other congenital cardiac anomalies. However, it may be seen isolated. Structures in LVOT such as tumor, vegetation, cysts may have attention for differential diagnosis. The number of cases is increasing with the routinely using of two-dimensional echocardiography.

Accessory mitral valve tissue is detected first early in children with symptoms of LVOT and is very rarely diagnosed in adults. One third of cases may asymptomatic, but commonly significant left ventricular outflow tract gradient can be detected in these cases, especially adult period. Optimal treatment of this anomaly is surgery if there is a significant LVOTO.

In this report, we presented the three asymptomatic adult cases with accessory mitral valve tissue, without increased gradient in LVOT. Surgical excision was recommended to the first case in another hospital with diagnosis of cardiac cyst. Two cases are presented.

Key Words:
Accessory mitral valve tissue, Asymptomatic adults, Normal LVOT gradient.

Introduction

Accessory mitral valve tissue (AMVT) is a congenital anomaly and may cause left ventricular outflow tract obstruction (LVOTO) rarely[1-3]. Although often associated with other cardiac anomalies, it might be seen as isolated. The first case of AMVT was described as early as 1842[4], and the first surgical management of this rare condition dates back to 1963[5]. The first report for this type of lesion in the echocardiography literature appeared in 1985[6].

Currently, there are approximately 100 cases of AMVT reported in the literature, less than 30 of which are described in the adult population[7]. Typically, AMVT presents early in life with symptoms of LVOT obstruction. In adults, reduced exercise capacity syncope, cerebrovascular and thromboembolic events and transient ischemic attack may be first symptom. There are no additional findings apart from murmur in approximately 30% of patients[8].

In this report, we presented three asymptomatic adult cases that diagnosed with isolated congenital accessory mitral valve tissue.

Case 1

Twenty five years old male patient was referred to our Clinic from the Cardiovascular Surgery Department for the reevaluation of patient before surgery. Transthoracic echocardiography (TTE) had been performed in another hospital because of murmur, and cystic structure in the ventricular side of anterior mitral leaflet had been detected, and surgical excision had been recommended.

There was no any sign on physical examination other than 2/6 systolic murmur on aortic origin. We performed transthoracic echocardiography (TTE), and detected a cyst-like structure, mobile, attached to the ventricular side of anterior mitral leaflet. This structure moved to LVOT during systole has not produced an increased gradient (Figure 1 A). Transesophageal echocardiography revealed an accessory mitral valve tissue adhering to the anterior mitral valve leaflet and ballooning into the left ventricular outflow tract during systole (Figure 1 B). In the absence of symptoms and relevant obstruction of the left ventricular outflow tract, the patient is being followed up without surgical intervention.

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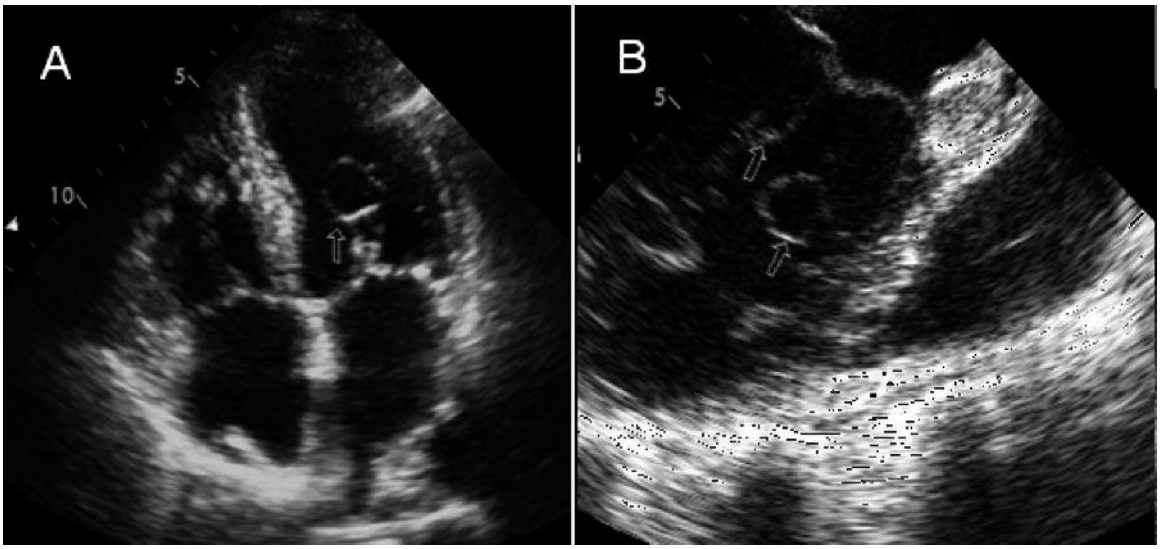

Figure 1. *A*, Echocardiographic view of the twenty five years old patient on apical four chamber view. The accessory mitral valve tissue was attached to anterior mitral leaflet. *B*, Accessory mitral valve tissue in TEE.

Case 2

Forty two years old female patient was referred to our Clinic due to 3/6 systolic ejection murmur on the second right intercostal area. The patient was asymptomatic and in good physical condition. There were no abnormal finding on electrocardiogram and chest X-Ray. TTE showed leaflet-like structure, attached anterior mitral leaflet, about 17 × 15 mm size. 12 mmHg gradient was estimated on LVOT. TEE was showed an accessory mitral valve tissue attached anterior mitral leaflet, and no associated other congenital anomalies. The patient was followed up without any surgical intervention.

Case 3

55 years old female patient was referred to our Clinic for the preoperative risk assessment. In the medical history, she had dyspnea with exertion, NYHA Class 2. She was noted to have a blood pressure of 140/90 mmHg, and a regular heart rate of 82 beats/min. Heart exam revealed a 3/6

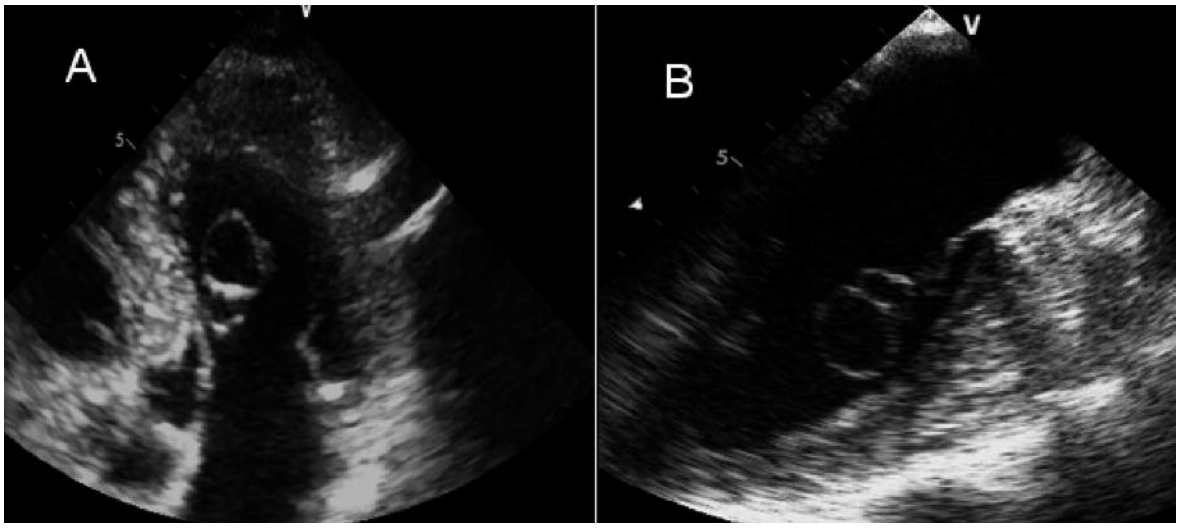

Figure 2. *A*, Echocardiographic view of AMVT obtained from apical four chamber. *B*, TEE showed attaching to anterior mitral leaflet of AMVT tissue.

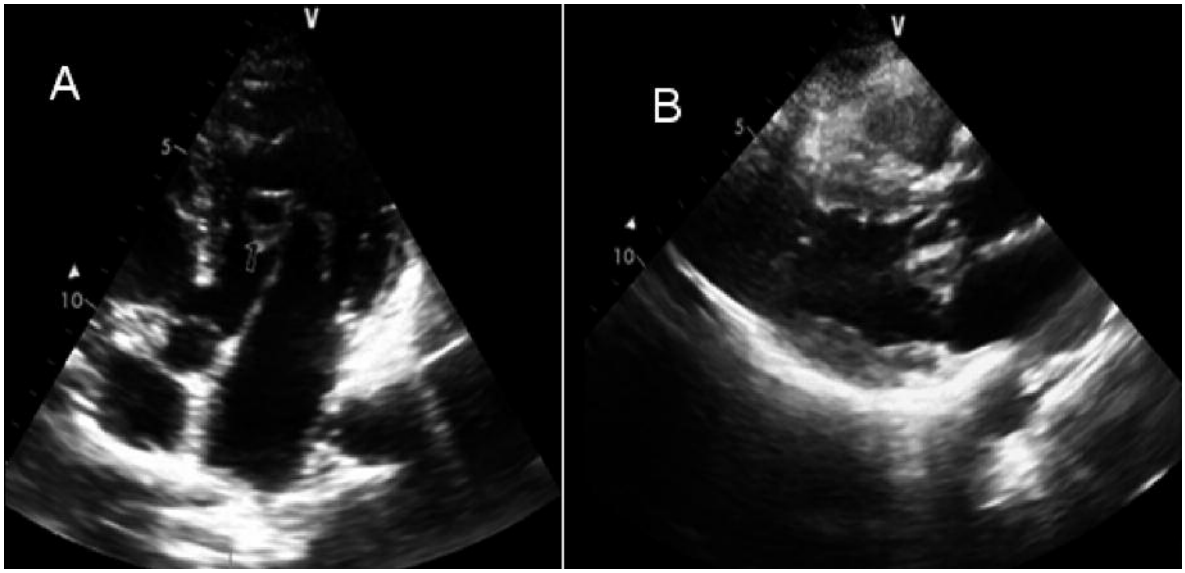

Figure 3. *A*, Echocardiographic images of 55 years old female patient from apical four chamber view. *B*, Parasternal long axis image, recorded during systole.

systolic ejection murmur at the aortic origin radiated to neck. Body mass index (BMI) was calculated as 33 kg/m^2. ECG was show normal sinus rhythm. Cardiothoracic ratio was estimated 0.55 on chest X-Ray. Echocardiography revealed an abnormal structure, compatible for accessory mitral valve, about 13 × 12 mm size. 15 mmHg gradient was estimated on LVOT. TEE was recommended to the patient, but did not accept. The patient was followed-up without other problems.

Discussion

AMVT is a rare congenital anomaly and its true incidence is unknown. AMVT is being reported every 26 000 echocardiographic examination in adult population[9]. Structures in LVOT such as tumor, vegetation, cysts may have attention for differential diagnosis.

Embryological formation of AMVT is not understood clearly. Abnormality in differentiation of mitral valve from endocardial cushion tissue during development of the heart may have lead to this anomaly[10]. AMVT is usually attaches on the anterior leaflet, corda tendineaes or papillary muscles.

Usually, there is an associated congenital anomaly in cases with AMVT. Moreover, majority of the cases may have symptoms of LVOTO such as angina and dyspnea with exercise and syncope in first decade[7]. One-third of the cases are asymptomatic and only murmur and mild LVOT gradient may occur. In literature, there are AMVT cases, without significant LVOT obstruction, caused cerebrovascular embolism[11].

LVOT obstruction in AMVT cases thought to be related with deplacement of accessory tissue to LVOT[2,12]. Excess tissue in the LVOT may cause to a significant gradient in some cases. However, reason of the LVOT gradient is progressive fibrosis of the tissue in majority of cases[13-15].

Differential diagnosis of subaortic stenosis due to AMVT and valvular aortic stenosis may be difficult in clinical examination. The number of cases is increasing with the routinely using of two-dimensional echocardiography. Structure and sizes, deplacement of the LVOT and severity of the LVOT obstruction can be evaluated with echocardiography[16,17]. Especially after the development on TEE images, differential diagnosis of AMVT to be a problem, rarely.

Surgical treatment is necessary in cases with AMVT in the presence of associated cardiac anomalies or symptoms of LVOTO. Additionally, surgery must be considered in the presence of findings of embolization, caused by AMVT[11]. Differentiation of AMVT from native mitral valve tissue is difficult in exploration[18]. Therefore, it is important to not overlook of the AMVT during preoperative echocardiography in patients with other congenital cardiac anomaly, candidate for surgery.

All of the presented cases were asymptomatic adults and there was no physical examination. Cranial computed tomography (CT) was performed in case 1 and 2 for exclusion of silent cerebral embolization. Case 3 did not accept the cranial CT. We recommended to patients echocardiography per two years, but not any medication such as infective endocarditic prophylaxis.

In the review of the literature, majority of the patients have additional cardiac anomalies. Moreover, most of them have symptoms due to significant gradient in LVOT. These cases are unique because of the lack of additional cardiac anomalies, any symptoms and significant LVOT gradient makes our cases unique.

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