ARCHIVES OF THE TURKISH SOCIETY OF CARDIOLOGY

Official journal of the TURKISH SOCIETY OF CARDIOLOGY

Accessory Mitral Valve Tissue in Both Obstructive and Nonobstructive Hypertrophic Cardiomyopathy Cases

Obstrüktif ve Nonobstrüktif Hipertrofik Kardiyomiyopati Vakalarında Aksesuar Mitral Kapak Dokusu

ABSTRACT

Accessory mitral valve tissue (AMVT) is a rare congenital cardiac anomaly often associated with other congenital heart defects. Typically diagnosed in childhood, its presence in adulthood is exceptionally rare, especially in patients with hypertrophic cardiomyopathy (HCM). The coexistence of these two conditions can complicate the diagnosis, particularly in patients with left ventricular outflow tract (LVOT) gradient formation. This case series highlights the importance of multimodal imaging in accurately identifying AMVT, differentiating it from other conditions, and determining the various morphologies of AMVT.

Keywords: Accessory mitral valve tissue, echocardiography, hypertrophic cardiomyopathy

ÖZET

Aksesuar mitral kapak dokusu, sıklıkla diğer konjenital kalp defektleriyle birlikte görülen nadir bir konjenital kalp anomalisidir. Tipik olarak daha erken yaşlarda teşhis edilmesine rağmen, özellikle hipertrofik kardiyomiyopati hastalarında erişkin dönemde görülmesi son derece nadirdir. Bu iki durumun bir arada bulunması, özellikle sol ventriküler çıkış yolu gradyanı oluşumu olan hastalarda tanıyı zorlaştırabilir. Bu vaka serilerinde, aksesuar mitral kapak dokusunun doğru bir şekilde tanımlanmasında, diğer durumlardan ayırt edilmesinde ve bunun farklı morfolojilerinin tanımlanmasında farklı görüntüleme tekniklerinin kullanımının önemini vurguluyoruz.

Anahtar Kelimeler: Aksesuar mitral kapak dokusu, ekokardiyografi, hipertrofik kardiyomiyopati

CASE REPORT
OLGU SUNUMU

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Received: January 03, 2024
Accepted: March 01, 2024

Cite this article as: Türkmen İ, Güler A, Atmaca S, Doğan İ, Babur Güler G. Accessory mitral valve tissue in both obstructive and nonobstructive hypertrophic cardiomyopathy cases. *Turk Kardiyol Dern Ars.* 2025;53(3):206-210.

DOI:10.5543/tkda.2024.15691

Accessory mitral valve tissue (AMVT) is an uncommon cardiac anomaly characterized by the presence of additional tissue near the mitral valve in the left atrium. This anomaly often coexists with other congenital heart defects such as ventricular septal defects (VSD), atrial septal defects (ASD), and tetralogy of Fallot.[1] Approximately 70% of cases of this condition are identified during childhood, with adult diagnoses being exceedingly rare.[2] However, there are reports of AMVT in patients with hypertrophic cardiomyopathy (HCM), though these are much less common.[3,4] The co-occurrence of these two genetic conditions can lead to diagnostic confusion. In fact, a left ventricular outflow tract (LVOT) gradient detected in patients with HCM is usually associated with systolic anterior motion (SAM), whereas the presence of AMVT can cause an LVOT gradient without SAM, presenting a challenge in differentiating between the two conditions. AMVT, like SAM, can extend to the septal region and cause gradient formation. This differentiation is particularly important in treatment planning for symptomatic patients. For this reason, it is critical to differentiate between patients with obstructive and nonobstructive HCM patterns.

Echocardiography, 3D echocardiography, and cardiac magnetic resonance imaging (MRI) are essential diagnostic tools for identifying AMVT.[5] Multimodal imaging is crucial for confirming the diagnosis and determining the appropriate treatment. In these two cases, we aim to emphasize the importance of recognizing different morphologies of AMVT coexisting with HCM and diagnosing them through multimodal imaging.

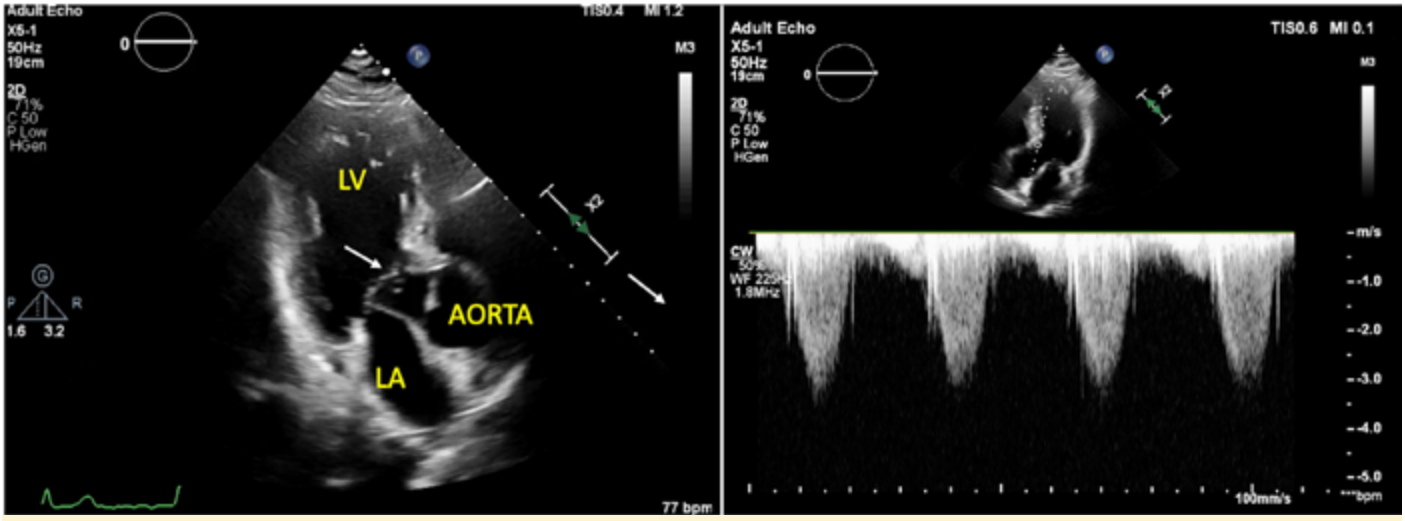

Figure 1. Mitral accessory tissue (white arrow) mimicking systolic anterior motion in transthoracic echocardiography apical 3-chamber view (A) and left ventricular outflow tract gradient with a maximum of 28 mmHg measured by continuous-wave Doppler (B) (LA, Left Atrium; LV, Left Ventricle).

Case Reports

Case 1

A 41-year-old male patient was admitted to our center with complaints of palpitation. His medical history was otherwise unremarkable. Electrocardiography (ECG) results displayed sinus rhythm and inverted T-waves attributable to left ventricular hypertrophy. Transthoracic echocardiography (TTE) revealed a wall thickness of the anterior septum of 18 mm, indicating asymmetric left ventricular hypertrophy.

The mitral valve leaflets appeared elongated, with the anterior leaflet protruding into the LVOT during systole, accompanied by SAM (Video 1). The length of the anterior leaflet was measured at 29 mm, indicating an increase, while the posterior leaflet measured 13 mm, which is within normal limits. A resting peak LVOT gradient of 13 mmHg was observed, which increased to 28 mmHg after the Valsalva maneuver (Figure 1).

Transesophageal echocardiography (TEE) was performed for a more detailed evaluation of the structure extending from the mitral valve to the LVOT. TEE showed that the accessory tissue on the mitral valve was directed towards the LVOT (Figure 2, Video 2). It was noted that this accessory tissue mimicked SAM and moved within the LVOT to create a systolic jet, despite the absence of actual SAM, as indicated by TTE. Specifically, transgastric views demonstrated that the accessory tissue caused LVOT narrowing without the presence of true SAM (Figure 3, Video 3).

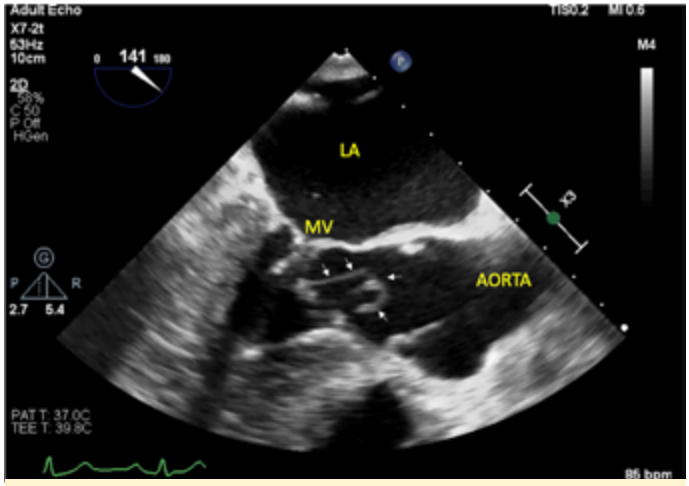

Figure 2. Accessory mitral valve tissue (white arrows) extending to the left ventricular outflow tract in transesophageal echocardiography midesophageal long axis view (LA, Left Atrium; MV, Mitral Valve).

To further analyze the tissue morphology, cardiac MRI was performed. The cardiac MRI (CMR) findings confirmed the asymmetric septal hypertrophy morphology and revealed an accessory tissue with a thickness of 2.4 mm causing a systolic jet flow in the LVOT and narrowing the LVOT, originating from below the anterior leaflet of the mitral valve (Figure 4, Video 4).

The presence of mitral accessory tissue without SAM was confirmed through multimodal imaging, integrating echocardiography and CMR findings.

Additionally, to establish a definitive diagnosis, genetic testing revealed a mutation in the myopalladin (MYPN) gene, which

ABBREVIATIONS

AMVT	Accessory mitral valve tissue
ASD	Atrial septal defect
ECG	Electrocardiography
HCM	Hypertrophic cardiomyopathy
ICD	Implantable cardioverter-defibrillator
LVOT	Left ventricular outflow tract
MRI	Magnetic resonance imaging
SAM	Systolic anterior motion
TEE	Transesophageal echocardiography
TTE	Transthoracic echocardiography
VSD	Ventricular septal defects

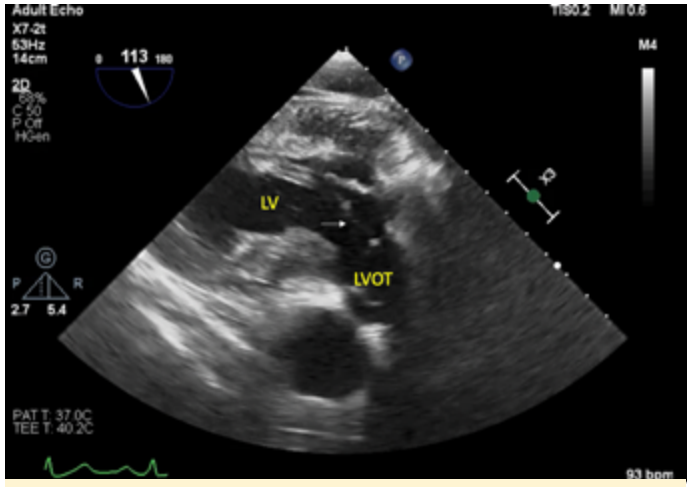

Figure 3. Accessory mitral valve tissue (white arrow) extending into the LVOT without systolic anterior motion transesophageal echocardiography transgastric view (LV, Left Ventricle; LVOT, Left Ventricular Outflow Tract).

is rarely detected. The patient's symptoms were effectively managed with bisoprolol treatment, thereby eliminating the need for surgical intervention. The patient is currently under observation and remains asymptomatic.

Case 2

A 47-year-old male patient was referred to our center for further evaluation and management with a previously confirmed diagnosis of HCM through genetic testing, which identified a mutation in the myosin-binding protein C-3 (MYBPC3) gene eight years prior. An implantable cardioverter-defibrillator (ICD) was implanted at that time. Physical examination revealed a 3/6 systolic murmur, but no other significant findings were present. The ECG displayed sinus rhythm and signs of left ventricular hypertrophy.

Initial TTE showed asymmetrical septal hypertrophy, SAM, and a resting gradient of 33 mmHg at the LVOT, which increased to 45 mmHg with the Valsalva maneuver and 84 mmHg post-exercise (Figure 5). The mitral anterior leaflet measured 25 mm, slightly increased, while the posterior leaflet was 13.6 mm, within normal limits. Additionally, a string-like abnormal structure was visualized in the LVOT (Video 5).

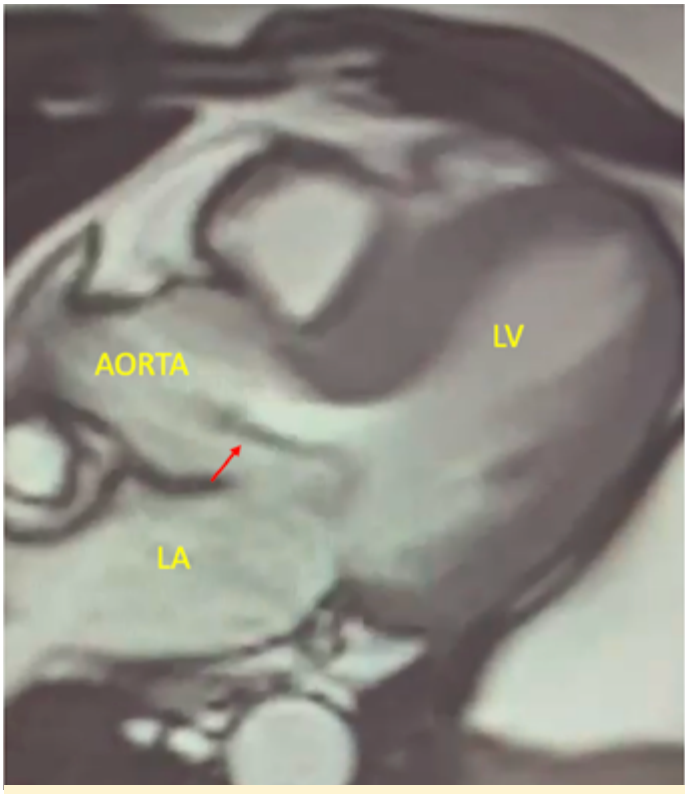

Figure 4. Cardiac MRI long axis projection of the 3-chamber view where accessory mitral valve tissue (red arrow) is seen extending from the anterior leaflet of the mitral valve to the LVOT (LV, Left Ventricle; LVOT, Left Ventricular Outflow Tract).

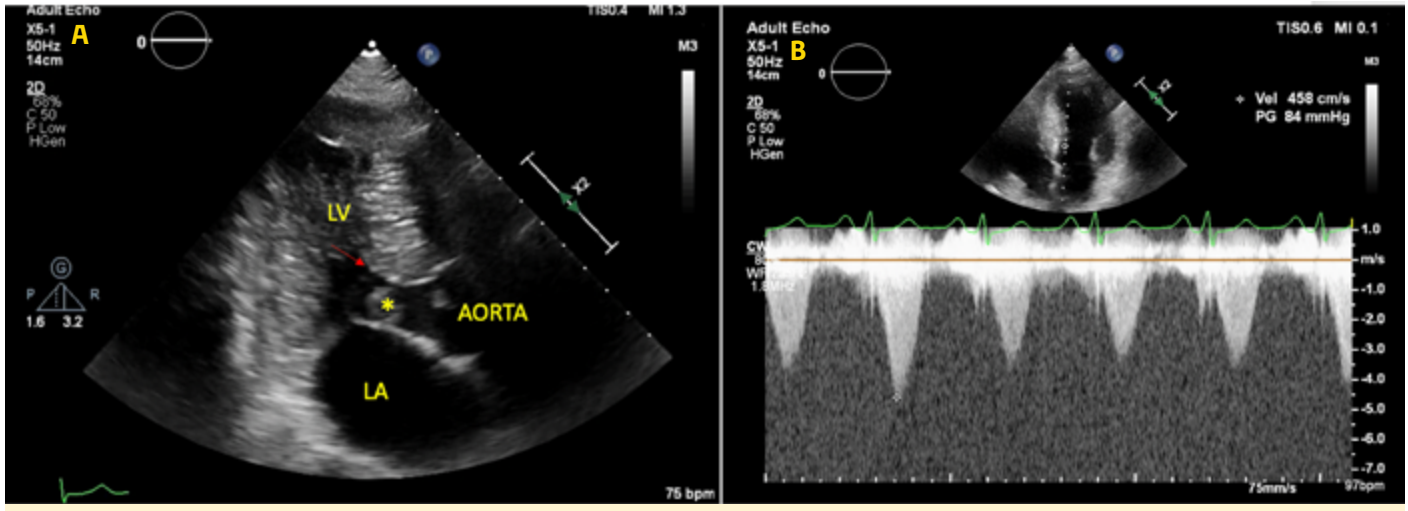

Figure 5. Accessory mitral valve tissue (*) and systolic anterior motion (red arrow) in transthoracic echocardiography apical 3-chamber view (A) and left ventricular outflow tract gradient with a maximum of 84 mmHg measured by continuous-wave Doppler (B) (LA, Left Atrium; LV, Left Ventricle).

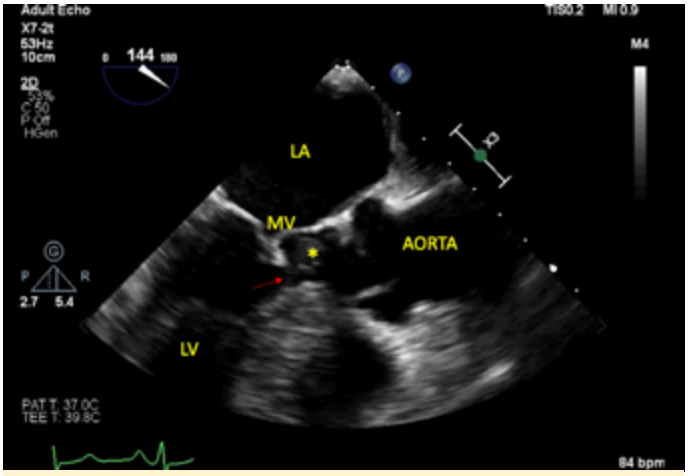

Figure 6. Accessory mitral valve tissue (*) extending to the left ventricular outflow tract with the presence of systolic anterior motion (red arrow) in transesophageal echocardiography midesophageal long axis view (LA, Left Atrium; MV, Mitral Valve).

TEE provided detailed insights into the mitral valve, showing SAM in both mitral valves and chordae. A circular and fibrillar structure measuring 1.34 x 0.4 cm, consistent with mitral accessory tissue, was observed beneath the anterior leaflet of the mitral valve (Figure 6). Moderate mitral regurgitation was detected from the mitral valve to the left atrium, with color Doppler showing a posterior eccentric and central regurgitation pattern. X-plane images and 3D imaging clearly depicted the accessory tissue extending to the LVOT (Videos 6, 7). Systolic blunting was also noted in the pulmonary veins. Due to the incompatible ICD lead, CMR was not feasible.

For symptom management, disopyramide was added to the existing metoprolol regimen. The patient has remained asymptomatic during the follow-up period.

Discussion

The objective of this case series is to report two cases of HCM featuring AMVT and to emphasize the heterogeneous clinical manifestations and diagnostic difficulties associated with this rare condition. The initial presentation in the first case was misleading, as AMVT was initially misinterpreted as obstructive HCM due to SAM detected on TTE. However, additional imaging modalities, including TEE and CMR, were utilized to confirm the presence of AMVT without SAM. The second case involved a patient with pre-existing SAM who was diagnosed with AMVT using multimodal imaging, including TEE and CMR. Although asymptomatic, both patients received conservative medical treatment, as surgical intervention was deemed unnecessary.

The prevalence of mitral accessory tissues, first described in 1842, is not well established because it is mostly detected incidentally during echocardiography and surgical procedures.[2,6] The incidence is higher in men. Rovner et al.[7] suggested that the incidence in the adult population is 1 in 26,000 echocardiographic procedures. Although the diagnosis is typically made in childhood, cases have been diagnosed as late as 77 years of age, according to the literature. It can be argued that the frequency of AMVT in adults is likely to increase with advancements in imaging techniques.[1]

The precise embryological mechanism of AMVT formation is unclear but is thought to result from abnormal or incomplete separation of the mitral valve from the endocardial cushions.[8] Prifiti et al.[1] have established a classification system for AMVT based on its mobility, resulting in two primary types: fixed and mobile. This classification is further refined based on insertion, relationship to the mitral valve, and chordae development. The type IIB leaflet form, situated on the ventricular side of the anterior mitral valve leaflet, is the most frequently encountered presentation. However, it is essential to recognize that other types of AMVT can also occur, each with unique features and implications for patient management. A comprehensive understanding of the various classifications and presentations of accessory mitral valve tissue is essential for accurate diagnosis and optimal treatment planning. AMVT is pathophysiologically linked to LVOT obstruction.[9] Two mechanisms have been identified as underlying the occlusion in patients with AMVT: (i) the mass effect of the accessory tissue and (ii) the progressive accumulation of fibrous tissue due to the turbulent flow created by AMVT.[2] While the majority of patients with mitral accessory tissue are asymptomatic, some may exhibit symptoms such as fatigue and dyspnea. Symptomatic presentation typically occurs when the mean gradient at the LVOT exceeds 50 mmHg.[10]

The two reported cases highlight the coexistence of genetically confirmed HCM and a congenital anomaly known as accessory mitral valve tissue. It is worth noting that elongated and redundant mitral valve leaflets are frequently observed in individuals with HCM, indicating a common morphological characteristic in these patients.[11] However, there are reports in the literature indicating the presence of mitral accessory tissue in HCM patients, which may be confused with SAM due to the LVOT gradient it generates.[3,12,13] Echocardiography remains the "gold standard" non-invasive method for evaluating AMVT. In our first case, the initial evaluation with TTE suggested SAM, leading to treatment being administered accordingly. However, further evaluation with TEE and CMR revealed that the primary issue was not SAM but rather the mass effect of the mitral accessory tissue at the LVOT. The second case presented with mitral accessory tissue and concomitant SAM, making it impossible to consider any symptomatic treatment for the patient independently of this mechanism. Both cases underscore the limitations of TTE alone and the necessity of advanced imaging techniques and multimodal imaging approaches in suspected patients.

Beta-blocker therapy may be considered for patients with AMVT who present with significant LVOT obstruction at rest or with provocation.[14] However, for symptomatic patients with AMVT and significant LVOT obstruction, surgical resection remains the primary cardiac intervention.[2]

Conclusion

AMVT coexisting with HCM is a rare condition that can cause LVOT obstruction and symptoms in affected patients. Multimodal imaging, including echocardiography, 3D echocardiography, and CMR, is critical in accurately identifying AMVT and differentiating it from other conditions, particularly HCM with SAM. Identifying the various morphologies of AMVT can have significant implications for treatment planning in symptomatic patients.

Informed Consent: Detailed written informed consent was obtained from the patient for the publication of this case report and accompanying images.

Peer-review: Externally peer-reviewed.

Author Contributions: Concept – İ.T., A.G., S.A., G.B.B.; Design – İ.T., A.G., S.A., G.B.B.; Supervision – G.B.B.; Resource – İ.T., A.G., S.A., İ.D.; Materials – S.A., İ.D.; Data Collection and/or Processing – A.G., İ.D.; Analysis and/or Interpretation – İ.D., G.B.G.; Literature Review – İ.T., A. G., S. A.; Writing – İ.T., A.G., S.A., G.B.B.;Critical Review – G.B.B.

Use of AI for Writing Assistance: Artificial intelligence assisted technologies were not used in this article.

Conflict of Interest: The authors have no conflicts of interest to declare.

Funding: The authors declared that this study received no financial support.

Video 1. Mitral accessory tissue mimicking systolic anterior motion in transthoracic echocardiography apical 3-chamber view.

Video 2. Accessory mitral valve tissue extending to the left ventricular outflow tract in transesophageal echocardiography midesophageal long axis view.

Video 3. Accessory mitral valve tissue extending into the LVOT without systolic anterior motion transesophageal echocardiography transgastric view.

Video 4. The Steady-State Free Precession (SSFP) cine sequence shows a long axis projection of the 3-chamber view where accessory mitral valve tissue extends from the anterior leaflet of the mitral valve to the left ventricular outflow tract, causing left ventricular outflow obstruction during systole.

Video 5. Accessory mitral valve tissue and systolic anterior motion in transthoracic echocardiography apical 3-chamber view.

Video 6. X-plane image of the left ventricular outflow tract and the presence of accessory mitral valve tissue in transesophageal echocardiography midesophageal long axis view.

Video 7. 3D transesophageal echocardiographic view of the left ventricular outflow tract, where the accessory tissue of the mitral valve extends.

References

1. Prifti E, Bonacchi M, Bartolozzi F, Frati G, Leacche M, Vanini V. Postoperative outcome in patients with accessory mitral valve tissue. *Med Sci Monit.* 2003;9(6):RA126–RA133.
2. Manganaro R, Zito C, Khandheria BK, et al. Accessory mitral valve tissue: An updated review of the literature. *Eur Heart J Cardiovasc Imaging.* 2014;15(5):489–497. [CrossRef]
3. Musumeci B, Spirito P, Parodi MI, Assenza GE, Autore C. Congenital accessory mitral valve tissue anomaly in a patient with genetically confirmed hypertrophic cardiomyopathy. *J Am Soc Echocardiogr.* 2011;24(5):592. [CrossRef]
4. Tennichi T, Taniguchi T. Accessory mitral valve tissue that caused a left ventricular outflow tract obstruction: A case report. *JA Clin Rep.* 2019;5(1):86. [CrossRef]
5. Buttar R, Baibhav B. Non-obstructive accessory mitral valve tissue in an asymptomatic adult: A case report and review of literature. *Cureus.* 2020;12(9):e10340. [CrossRef]
6. Sono J, McKay R, Arnold RM. Accessory mitral valve leaflet causing aortic regurgitation and left ventricular outflow tract obstruction. Case report and review of published reports. *Br Heart J.* 1988;59(4):491–497. [CrossRef]
7. Rovner A, Thanigaraj S, Perez JE. Accessory mitral valve in an adult population: The role of echocardiography in diagnosis and management. *J Am Soc Echocardiogr.* 2005;18(5):494–498. [CrossRef]
8. Yuan SM, Shinfeld A, Mishaly D, Haizler R, Ghosh P, Raanani E. Accessory mitral valve tissue: A case report and an updated review of literature. *J Card Surg.* 2008;23(6):769–772. [CrossRef]
9. Yetkin E, Cuglan B, Turhan H, Yalta K. Accessory mitral valve tissue: Anatomical and clinical perspectives. *Cardiovasc Pathol.* 2021;50:107277. [CrossRef]
10. Mardenli M, Samman A, Alkanj H, Babi A. Obstructive accessory mitral valve tissue in an adult patient: A case report. *J Med Case Rep.* 2019;13(1):184. [CrossRef]
11. Kaple RK, Murphy RT, DiPaola LM, et al. Mitral valve abnormalities in hypertrophic cardiomyopathy: Echocardiographic features and surgical outcomes. *Ann Thorac Surg.* 2008;85(5):1527–1535. [CrossRef]
12. Panduranga P, Eapen T, Al-Maskari S, Al-Farqani A. Accessory mitral valve tissue causing severe left ventricular outflow tract obstruction in a post-Senning patient with transposition of the great arteries. *Heart Int.* 2011;6(1):e6. [CrossRef]
13. Li Y, Hu Y, Wang J, Liu L. A rare case of accessory mitral valve tissue causing left ventricular outflow tract obstruction associated with parachute mitral valve, ventricular septal defect, bicuspid aortic valve, unruptured aneurysm of aortic sinus: A case report. *Eur Heart J Case Rep.* 2018;2(3):yty082. [CrossRef]
14. Al-Atta A, Khan H, Sosin M. Accessory mitral valve tissue causing features of left ventricular outflow tract obstruction- A case report and updated literature review. *J Ayub Med Coll Abbottabad.* 2019;31(2):276–278.