

Asymptomatic Accessory Mitral Valve Leaflet with Left Ventricular Outflow Tract Obstruction

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Abstract

Background: An accessory mitral valve leaflet is a rare congenital anomaly usually associated with left ventricular outflow tract (LVOT) obstruction. Fewer than 100 cases have been reported mostly occurring in children.

Case: An asymptomatic 43-year-old female who came in for pre employment clearance was noted to have a systolic murmur. Echocardiography revealed an accessory mitral valve tissue attached to the anterior mitral valve leaflet. There was also a significant gradient noted across the LVOT. Patient is presently asymptomatic and she was advised follow up for serial echocardiographic monitoring.

Diagnostics: Twelve lead electrocardiogram showed normal sinus rhythm with normal axis, poor R wave progression and no chamber enlargement. Chest

radiography was normal. 2D Echo revealed an echogenic, parachute-like structure billowing into the LVOT during systole. Interrogation of the said structure using Doppler revealed a significant pressure gradient of 45mmHg along the LVOT.

Conclusion: Accessory mitral valve with LVOT obstruction is an unusual finding. Clinical and echocardiographic recognition as well as surveillance is warranted in these patients. This is also important for determination of definitive management.

Keywords: accessory mitral valve leaflet, left ventricular outflow tract obstruction

Introduction

Accessory mitral valve leaflet is an uncommon entity. Fewer than 100 cases have been reported mostly in children.¹ They can be associated with other congenital anomalies such as transposition of the great arteries. These patients usually present with symptoms of left ventricular obstruction early in life. Early diagnosis is associated with improved outcomes after surgical treatment.² Asymptomatic cases, especially in the adult are rare and usually discovered as incidental findings on imaging examinations. We report a case of an asymptomatic woman with echocardiographic diagnosis of an accessory mitral valve leaflet.

Case

A 43-year-old female non-hypertensive and non-diabetic with good functional capacity was referred for clearance prior to employment. She had just given birth

three months prior via caesarian section. Otherwise, past medical and family history were unremarkable. Blood pressure was 100/60 with a heart rate of 76 beats per minute. Cardiovascular examination revealed a 2/6 mid systolic murmur at the left sternal border best heard at the base with no radiation to the neck. She had normal 1st and 2nd heart sounds. Electrocardiogram showed sinus rhythm, poor R wave progression with no hypertrophy. Chest xray findings were normal (Figure 1).

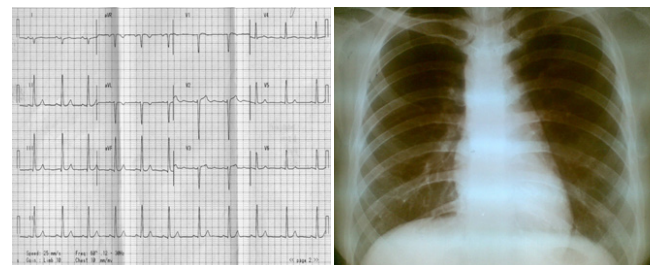


Figure 1. 12-L ECG showed a regular sinus rhythm with normal QRS axis. There was note of poor R-wave progression but no signs of any chamber enlargement or ischemia. (left) Chest X ray is normal (right)

Transthoracic two dimensional echocardiogram showed a structure attached to the anterior mitral valve leaflet which seems to be obstructing the left ventricular outflow tract (LVOT) on systole (Figure 2). There was note of an accessory mitral leaflet with chordal attachment to the

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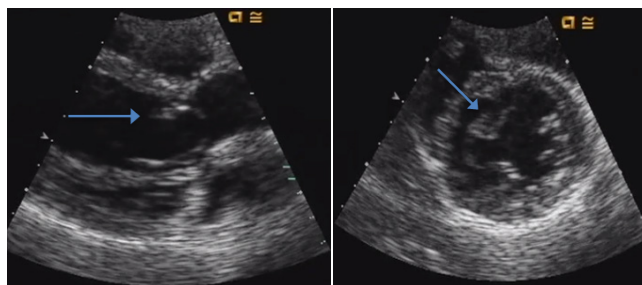


Figure 2. Parasternal long axis view in systole shows a mobile suspicious membrane seemingly obstructing the left ventricular outflow tract. (arrow, left) Short axis view at the level of the mitral valve shows the elongated density to be in direct contact with the ventricular septum and seemingly attached to the anterior mitral valve leaflet. (arrow, right)



Figure 3. Apical 4 chamber view shows an accessory mitral valve leaflet which is in contact with the ventricular septum with its chordae attached to the anterior papillary muscle (arrow, left) Doppler interrogation shows a flow turbulence proximal to the structure at the left ventricular outflow tract measuring around 45mmHg (right)

anterior mitral valve leaflet. Doppler interrogation of the LVOT showed dynamic pressure gradient with the highest pressure recorded at around 45mmHg (Figure 3). There was mild aortic regurgitation and mild mitral regurgitation. No other abnormalities were noted. The left ventricle was normal in size with no hypertrophy. Ejection fraction was 65% with adequate wall motion and contractility. Patient was then advised follow up with trans esophageal echocardiogram and serial monitoring.

Discussion

An accessory mitral valve is a rare congenital abnormality which is commonly associated with LVOT obstruction. Rovner A. et al. reported an incidence of one case per 26,000 examinations.³ Its incidence and natural history are not fully understood. It was hypothesized that its embryology may involve an abnormal or incomplete separation of the mitral valve from the endocardial cushion during cardiac development.⁴

Anatomically and based on intraoperative descriptions this anomaly has two classifications. Type I is fixed. This is further subdivided into IA if it is nodular and IB if its membranous. Type II is mobile and is further subdivided into IIA if pedunculated and IIB for leaflet like. The most common location is the anterior mitral leaflet.⁵ From the appearance on imaging our patient may have a type IIB.

Accessory mitral valves may be asymptomatic or may present with dynamic LVOT obstruction, usually simulating systolic anterior motion of the leaflet and a mass effect causing subaortic obstruction. Continued turbulence causes permanent deposition of fibrous tissue in the left ventricular outflow tract. The obstruction will result in pressure overload and hypertrophy similar in physiology to valvular aortic stenosis and hypertrophic cardiomyopathy. Chronic pressure overload can lead to subendocardial ischemia and eventual heart failure.⁶ Prifti et al. reported different findings in 90 patients.⁷ Diagnosis was mostly made in the first decade of life accompanied by significant LVOT obstruction. 15 patients had evidence of mild LVOT obstruction and three had no significant pressure gradient. Our patient has evidence of dynamic LVOT obstruction derived by Doppler measurements. Manganero et al.⁸ noted a prevalence of 86.6% for the obstructive form with 37.5% presenting with no symptoms and less than 30% presenting with dyspnea and heart failure.

Echocardiography is considered the optimum imaging of choice in the diagnosis. They can be mistaken for other lesions such as tumors, masses or even an elongated chordae. Transthoracic echo is usually adequate but transesophageal echocardiogram adds more diagnostic value and will enable proper clarification of its nature as well as its morphology and anatomic characteristics.⁹ These modalities enable a more non invasive approach. Our patient only presented with an asymptomatic murmur. According to Jiang et al the presence of dynamic LVOT obstruction with significant pressure gradient (defined as mean gradient of ≥ 25 mmHg) may require surgery even if the patient is asymptomatic and has been associated with favorable post operative and long term outcomes.¹⁰ We recommend close follow up as well as transesophageal echocardiogram to determine the anatomy and significant chordal attachments. After echocardiographic recognition we advise serial monitoring for progression.

Conclusion

Accessory mitral valve leaflet with LVOT obstruction is a rare finding. The increasing use of echocardiography has enabled identification of these otherwise unusual lesions as well as monitoring for potential complications. This case emphasizes the importance of proper clinical and echocardiographic recognition enabling clinicians to facilitate options for definitive management.

References

1. Roza, et al. Accessory Mitral Valve without Left Ventricular Outflow Tract Obstruction in an Adult, Tex Heart Inst J. 2008; 35(3): 324–326.
2. Tanaka H, Okada K, Yamshita T, Nakagiri K, Matsumori M,

- Okita Y.** Accessory mitral valve causing left ventricular outflow tract obstruction and mitral insufficiency. *J Thorac Cardiovasc Surg* 2006;132(1):160–1.
3. **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):494–8
 4. **Aoka Y, Ishizuka N, Sakomura Y, Nagashima H, Kawana M, Kawai A, Kasanuki H.** Accessory mitral valve tissue causing severe left ventricular outflow tract obstruction in an adult. *Ann Thorac Surg* 2004;77(2):713–5
 5. **Prifti E, Frati G, Bonacchi M, Vanini V, Chauvaud S.** Accessory mitral valve tissue causing left ventricular outflow tract obstruction: case reports and literature review. *J Heart Valve Dis* 2001;10(6):774–8.
 6. **Tanaka H, Kawai H, Tatsumi K, Kataoka T, Onishi T, Yokoyama M, Okita Y.** Accessory mitral valve associated with aortic and mitral regurgitation and left ventricular outflow tract obstruction in an elderly patient: a case report. *J Cardiol* 2007; 50(1):65–70.
 7. **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):RA126–33.
 8. **Manganero et al,** Accessory Mitral Valve Tissue, an updated review of the literature, *European Heart Journal - Cardiovascular Imaging* May 2014,15(5)489-497;DOI: 10.1093/ehjci/jet163
 9. **Eiriksson H, Midgley FM, Karr SS, Martin GR.** Role of echocardiography in the diagnosis and surgical management of accessory mitral valve tissue causing left ventricular outflow tract obstruction. *J Am Soc Echocardiogr* 1995;8(1):105–7
 10. **Jiang S, Zhang T, Sheng W, Gao C,** Accessory mitral valve causing left ventricular outflow tract obstruction, *Heart Surg Forum*. 2010 Aug;13(4):E267-8. doi: 10.1532/HSF98.20091190