An Unusual Etiology of Presyncopal Episodes: Dynamic Left Ventricular Outflow Tract Obstruction Secondary to an Elongated Anterior Mitral Valve Leaflet

Dynamic left ventricular outflow tract obstruction (LVOTO) refers to the restriction of blood flow from the left ventricle (LV) to the aortic arch. LVOTO can be classified as valvular, sub-valvular, or supravalvular.¹ Hemodynamically, it is defined by an instantaneous peak pressure gradient (PPG) at the LV outflow of \geq 30 mmHg, either at rest or upon provocation.^{1, 2} Gradients \geq 50 mmHg, whether at rest or provoked, are considered clinically significant, as they are associated with symptom development and serve as the threshold for initiating standard therapy.^{2, 3} In refractory cases, advanced pharmacological or invasive interventions might be required.³ Although LVOTO is most commonly described in patients with hypertrophic cardiomyopathy (HCM), it can also be congenital or secondary to other conditions, such as bicuspid aortic valve with coarctation of the aorta and cardiac amyloidosis.^{2, 3}

A 45-year-old man with a medical history of dyslipidemia presented with presyncopal episodes occurring during exertion over the past 18 months. He reported near-fainting episodes lasting for seconds to several minutes during physical activity, accompanied by generalized fatigue, lightheadedness, dizziness, and rarely, blurry vision. The patient was on atorvastatin 20 mg daily (Viatris Inc., USA) for dyslipidemia. Clinical examination, including supine and standing blood pressure and heart rate measurements, lung auscultation, palpitation of the peripheral pulses, and neurological examination

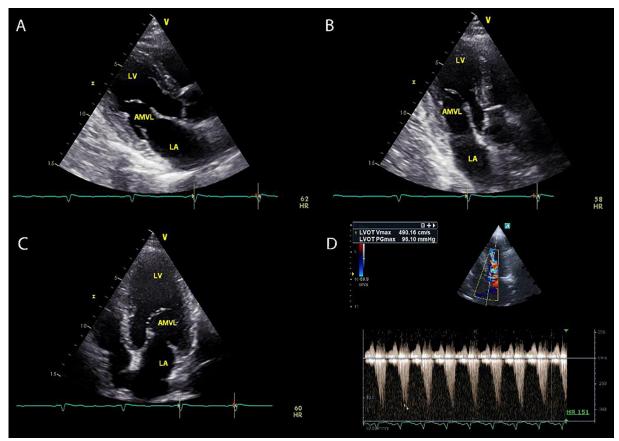


Figure 1: The transthoracic echocardiogram (TTE) shows an extremely elongated anterior mitral valve leaflet (AMVL). A: Parasternal long axis view; B: Apical 3-chambers view; C: Apical 4-chambers view. D: Dynamic obstruction of the LVOT during DSE; AMLV: Anterior mitral valve leaflet; TTE: Transthoracic echocardiogram; LVOT: Left ventricular outflow tract; LA: Left atrium; LV: Left ventricle; DSE: Dobutamine stress echocardiogram

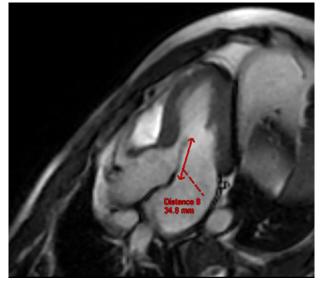
Copyright: ©Iranian Journal of Medical Sciences. This is an open-access article distributed under the terms of the Creative Commons Attribution-NoDerivatives 4.0 International License. This license allows reusers to copy and distribute the material in any medium or format in unadapted form only, and only so long as attribution is given to the creator. The license allows for commercial use. was unremarkable except for a 2/6 systolic murmur best heard at the apex. Initial cardiological evaluations, including an electrocardiogram and 48-hour Holter monitoring, revealed no abnormalities. Transthoracic echocardiogram (TTE) (GE Vivid 9, USA, and Alpinion ECUBE 8, South Korea) demonstrated preserved LV contractility (ejection fraction: 65%), normal cardiac chamber dimensions, and no evidence of hypertrophy. However, an elongated anterior mitral valve leaflet (AMVL) with mild mitral valve regurgitation (MR) was observed (figure 1 A-C).

Laboratory investigations, including complete blood count (cbc), erythrocyte sedimentation rate (ESR), C-reactive protein, thyroid indices, electrolytes, blood gas analysis, troponin, D-dimer concentrations, and cortisol levels were all within normal limits. Computed tomography (CT) scans of the chest and abdomen (North Star Imaging Inc., USA) and magnetic resonance imaging (MRI) of the brain (Philips Ingenia MRI 3T, Germany) revealed no abnormalities. An exercise stress test was performed, which showed no evidence of ischemia. However, the test was terminated prematurely due to the patient experiencing near-fainting episodes accompanied by dizziness and lightheadedness. Subsequently, a dobutamine stress echocardiogram (DSE) was conducted, which also revealed no signs of ischemia but reproduced the same symptoms. During the DSE, dynamic LVOTO was identified, caused by the motion of the AMVL, with a PPG of 96 mmHg (figure 1D). A cardiac MRI scan (GE SIGNA EXCITE II, USA) confirmed the presence of an elongated AMVL (34.8 mm, normal range: 22-23 mm) with no other additional abnormalities (figure 2). The patient was initiated on bisoprolol (Merck KGaA, Darmstadt, Germany) 5 mg daily, which resulted in significant symptomatic improvement.

To our knowledge, this study is the first published case of dynamic LVOT obstruction caused by an abnormally elongated AMVL leading to presyncopal episodes in a patient without HCM or any other obvious cardiac pathology. Although most cases of exercise-induced syncope or presyncope are of neurocardiogenic origin and are benign, persistent symptoms warrant the exclusion of structural cardiovascular abnormalities and/or cardiac arrhythmias.⁴

Several studies suggested that an abnormally elongated AMVL was closely related to the severity of LVOTO in patients with HCM.^{5,6} However, the flow dynamics and drag forces generated by an abnormally elongated AMVL can interact with the interventricular septum, leading to systolic anterior motion (SAM) of the MV. This mechanism can result in LVOTO and MR even in the absence of cardiac hypertrophy, potentially precipitating presyncopal episodes.^{5,6} Several diagnostic maneuvers, such as the Valsalva maneuver and exercise testing combined with simultaneous echocardiography, were employed to diagnose LVOTO.^{3,7} Although DSE is not routinely used to identify latent LVOTO, it was instrumental in this case for establishing a definitive diagnosis.^{4,7} Additionally, the potential association between syncope or presyncope and occult malignancy (particularly lung and brain cancers) or paraneoplastic phenomena (e.g., carcinoid syndrome or pheochromocytoma) prompted further investigation with CT scans of the chest and abdomen, as well as MRI of the brain in this patient.⁸

Figure 2: Cardiac MRI (3-chamber balanced SSFP cine images) demonstrates an elongated anterior mitral valve leaflet (AMVL) measuring 34.8 mm, exceeding the normal range of 22–23 mm. AMLV: Anterior mitral valve leaflet; SSFP: Steady-state free precession; MRI: Magnetic resonance imaging



Written informed consent was obtained from the patient for the publication of all data included in this report, with the condition of maintaining anonymity.

Authors' Contribution

All authors have participated sufficiently and equally in the preparation and submission of this manuscript to take public responsibility for its content. Specifically, to the: (i) conception and design of the work and interpretation of data for the work; (ii) drafting the work and reviewing it critically for important intellectual content; (iii) final approval of the version to be published, and (iv) agreement to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work were appropriately investigated and resolved.

Conflict of Interest: None declared.

Keywords • Dynamic left ventricular outflow tract obstruction • Presyncope • Cardiomyopathy, Hypertrophic • Magnetic resonance imaging

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