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A case report of chordal systolic anterior motion without hypertrophic cardiomyopathy misdiagnosed with acute coronary syndrome

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Case Report

Abstract

Chordal systolic anterior motion (SAM) refers to the anterior motion of the mitral valve towards the left ventricular outflow tract (LVOT) that occurs during systole. Chordal SAM is a potentially fatal condition. Chordal SAM is linked to an elevated risk of sudden death and can lead to significant obstruction of the left ventricular outflow tract (LVOT) and mitral regurgitation. It is crucial to acknowledge and consider this phenomenon when assessing individuals experiencing dyspnea and chest discomfort. By doing so, unnecessary therapies related to heart failure and heart attack might be avoided. Ensuring timely diagnosis is crucial in order to prevent unnecessary and potentially dangerous therapies that can aggravate LVOT obstruction and lead to hemodynamic instability. In this paper, we present a 47-year-old woman who experienced sudden chest pain and dyspnea. The patient had a history of hypertension and previous myocardial infarction. ST-elevation was observed in the first ECG. The primary treatment for acute coronary syndrome was initiated. Coronary angiography demonstrated that coronary vessels were not obstructed. The diagnosis of chordal SAM was initially overlooked during the echocardiography, but it was later identified during a subsequent echocardiography after cardiac catheterization. When using beta-blockers, her clinical condition improved.

Keywords: Chordal Systolic Anterior Motion; Valvular Systolic Anterior Motion; Diuretic; LVOT Obstruction; Hypertrophic Cardiomyopathy

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Introduction

Systolic anterior motion (SAM) of the mitral valve was initially believed to be unique to hypertrophic cardiomyopathy (HCM), but it is now understood that it can occur in any situation that alters the dynamic complexity of the left ventricle. Despite initial findings that left ventricular outflow tract (LVOT) obstruction was present in every case of SAM, its manifestation has since been recognized to encompass a spectrum of symptoms, ranging from clinically asymptomatic to severe LVOT obstruction accompanied by hemodynamic compromise[1].

Echocardiography referrals for Japanese patients indicate a prevalence of 0.3% for SAM in the absence of HCM. Non-HCM patients exhibit a lower LVOT gradient, are elderly, and have a greater prevalence of sigmoid septum in comparison to HCM patients, among other clinical characteristics. SAM is distinguished from HCM patients by the presence of robust clinical symptoms, an elevated LVOT gradient, anterior mitral leaflet elongation, and mitral regurgitation[2]. This case report details the

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clinical presentation and challenging diagnosis of a patient who presented to the hospital complaining of chest pain. Upon investigation, it was determined that the patient had SAM with LVOT obstruction, but no HCM.

Case presentation

A 47-year-old woman was admitted to the emergency department after complaining of dyspnea and typical chest pain with a heaviness quality that radiated to her back. She declared that the quality and pattern of the chest pain were similar to those she experienced during her prior myocardial infarction (MI). The past medical history of the patient was significant for hypertension, dyslipidemia, and MI three years ago, following four angiographies and two percutaneous coronary interventions (PCI). Aspirin 80 mg daily, atorvastatin 40 mg daily, clopidogrel 75 mg daily, and diltiazem 40 mg BID were recorded as her drug history. Her electrocardiogram (ECG) revealed diffuse ST-segment depression and aVR ST-segment elevation, signifying a substantial change in comparison to her previous ECGs. On her prior angiography, no significant stenosis of the left main coronary artery was detected. Before angiography, initial echocardiography in the emergency department revealed mitral regurgitation with an EF of 45%.

We decided to perform an angiography due to the ECG alteration and the previous MI. According to the results of our angiography, there was no notable lesion, with patent stents of the right coronary artery and the left anterior descending artery. Upon admission, she was placed in the coronary care unit (CCU). We continued the slow infusion of nitroglycerin because of her persistent chest distress. Throughout her hospitalization, she developed severe dyspnea accompanied by a crackle sound on pulmonary auscultation. Frusemide was administered intravenously based on suspicion of decompensated heart failure. A second echocardiography examination was performed for reevaluation; the results revealed an estimated ejection fraction (EF) of 45% along with hypokinesia affecting the mid-anteroseptal, mid-inferoseptal, and base to mid-oblique walls. There was no evidence of LVH. The thickness at the base of the septum was 13 mm. It is noteworthy that we observed a sigmoid-shaped septum, a thickened mitral valve (MV), substantial chordal and valvular systolic anterior motion (SAM), and severe mitral regurgitation. Additionally, moderate degenerative mitral stenosis accompanied by moderate to severe calcification of the mitral annulus was observed. In addition, a substantial SAM and severe LVOT obstruction due to a sigmoid-shaped, hypercontractile base septum were evident. The LVOT gradient in tachycardia and after activity was 118 mmHg, while the LVOT gradient at rest was 67 mmHg. A 6 mmHg mean gradient was detected. Mild to moderate tricuspid regurgitation, a thickened aortic valve, and a mild PI were observed. Following the abovementioned results, nitroglycerin and lasix were discontinued, and volume-loading treatment with the addition of bisoprolol was prescribed. Her symptoms were alleviated after this treatment. She met the surgical criteria for mitral valve replacement. However, she declined to undergo the surgery and was discharged with a prescription for bisoprolol.

Discussion

The pathophysiology that results in SAM is not entirely comprehended. Research has demonstrated that SAM is linked to the imbalance between the length of the anterior and posterior leaflets. SAM causes mitral regurgitation and LVOT obstruction[3,4]. MI is another mechanism that is associated with SAM. SAM is rarely detected subsequent to MI. This is believed to be the result of changes in left ventricular geometry caused by the contrast between hypokinetic and hyperkinetic regions following acute MI. This results in a decreased LVOT and causes SAM. Inotropes and vasodilators, which are conventionally prescribed for cardiogenic shock, may exacerbate SAM and induce additional LVOT obstruction. The careful administration of beta-blockers is necessary for these patients[1].

Numerous hypotheses have been advanced regarding the cause of chordal SAM. Chordal anomalies, mitral annulus disruption, and congenital anomalies are frequently proposed as the causes of chordal SAM. However, chordal SAM can also be caused by systemic diseases such as diabetes, hypertension, and MI[1]. Chordal SAM in our patient is likely the result of prior MI and hypertension. In addition, mitral annulus calcification was detected by echocardiography in our patient, which may provide an additional explanation for chordal SAM.

Although most patients with SAM are attributed to HCM, SAM has become increasingly detected in patients without HCM. Aging and a sigmoid septum are clinical characteristics of chordal SAM in the absence of HCM. Patients with a sigmoid septum who have SAM and do not have HCM exhibit SAM not only under hyperkinetic conditions but also at rest[2]. Notably, a sigmoid-shaped septum was also identified in our patient, contributing to LVOT obstruction.

The majority of our understanding of the pathophysiology of chordal SAM is limited to patients with HCM. The pathophysiology of SAM in patients without HCM remains unclear, and our understanding is restricted to a few associations that are founded on a small population. Patients with SAM but no HCM demonstrate a lower LVOT gradient pressure and older ages in comparison to those with HCM. Nevertheless, certain factors, including anemia, anesthesia, acute MI, and mitral valve surgery, can elevate the LVOT gradient pressure. The LVOT gradient pressure can also be influenced by the morphology of SAM. For example, valvular SAM is linked to elevated gradient pressure in LVOT, increased mitral regurgitation, and a greater number of symptoms in comparison to chordal SAM. In addition, patients with SAM who do not have HCM are observed to have LVOT obstruction due to mitral valve elongation[2-4].

Recent research has demonstrated that conservative medical treatment is the preferable approach for SAM. The objective of this approach is to decrease the contractility of cardiac muscle and pulse rate by administering beta-blockers and increasing intravascular volume with fluids. The prescription of vasoconstrictors to enhance vascular resistance can also be beneficial[3]. Therefore, the principal treatments for patients with SAM consist of volume loading and beta-blockers. The treatment for SAM begins with volume loading. When this fails, a beta-blocker is used. In the event that SAM persists, surgical intervention is recommended[5-7]. Nonetheless, this method requires a diagnosis of chordal SAM. It can be difficult to diagnose chordal SAM in daily practice. Chordal SAM may cause LVOT obstruction. LVOT obstruction can result in elevated intraventricular pressures, increased cardiac workload, and myocardial ischemia. Due to these complications, symptoms such as syncope, chest discomfort, exercise intolerance, and heart failure might develop[8]. Due to the low prevalence of SAM, the diagnosis of SAM may be postponed, which may subsequently lead to complications of the condition. SAM is not a benign condition, and LVOT obstruction is potentially fatal[1,9]. These symptoms remind us that chordal SAM has the potential to resemble symptoms of ischemia and may direct our attention to an alternative diagnosis, such as coronary artery disease. Therefore, it is crucial that we incorporate SAM into our clinical approach, particularly when coronary angiography reveals no obstruction in the patient. The low prevalence of LVOT obstruction in chordal SAM further complicates the clinical approach and the diagnosis of this condition. Previously, it was believed that LVOT obstruction in chordal SAM without HCM was uncommon[10]. A recent study, however, revealed that LVOT obstruction is not uncommon in this population. LVOT obstruction in SAM without HCM can occur at rest and is not restricted to exercise alone, which can further complicate the situation[2].

Transcatheter treatment is an additional therapeutic approach to SAM. Transcatheter treatment involves the merging of the anterior and posterior leaflets to remove the leaflet from the LVOT. This invasive treatment simultaneously reduces regurgitation and the obstruction of the LVOT[4]. Nevertheless, recent research has demonstrated that the majority of patients experience advantages from conservative medical management of SAM, instead of invasive treatments[3].

Additionally, there are case reports that present patients who have SAM without HCM. In two of them, the patient had no prior medical history[11,12]. In the other, the patient had a significant medical history, including congenital heart disease and cardiac surgery[13]. The efficacy of treatment with beta-blockers and fluid resuscitation, particularly in emergency patients, is a commonality between our case and these case reports[11,12]. Nevertheless, the evidence is controversial in certain instances, particularly in the context of congenital diseases and previous cardiac surgery, and it is unclear whether to opt for medical therapy or surgery[13].

Conclusion

Chordal SAM is the anterior mitral valve motion that has the potential to obstruct the LVOT. It is critical

to recognize and take into account this phenomenon when diagnosing patients with dyspnea and chest pain; doing so can prevent the need for irrelevant treatments associated with heart failure and myocardial infarction. Preventing irrelevant harmful treatments, which can exacerbate LVOT gradients and cause hemodynamic worsening, depends on a prompt diagnosis. Patients who are diagnosed with ACS and undergo angiography without manifesting any obstruction during the procedure should be duly considered. The rarity of SAM, particularly in patients without HCM, complicates the diagnostic process. Despite historical limitations, our understanding of SAM has advanced. Consequently, it is critical to incorporate this phenomenon into diagnostic considerations.

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Conflict of interests

All authors declare that they have no conflicts of interest.

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Author's Contributions

MHA: Data curation; formal analysis; investigation; methodology; writing – original draft; review and editing. FO: Data curation; formal analysis; investigation; methodology; writing – original draft; review and editing. RHB: Data curation; formal analysis; investigation; methodology; writing – original draft.

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