A handicap in Takotsubo cardiomyopathy: dynamic outflow obstruction

Takotsubo kardiyomiyopatisinde bir handikap: Dinamik çıkım yolu obstrüksiyonu

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Summary– Takotsubo cardiomyopathy (TCM) is an unusual form of acute cardiomyopathy showing left ventricular apical ballooning. Patients with hypotension should undergo urgent echocardiography to determine if left ventricular outflow tract (LVOT) obstruction is present. This complication has been described in 10–25% of all TCM patients. That some patients do not survive an acute TCM event underscores the importance of prompt recognition and targeted management of dynamic LVOT obstruction. Described in the present report is the case of a TCM patient with cardiogenic shock, the hemodynamics of which worsened after inotropic agents were administered, alleviated after serum fluid and beta-blocker treatment.

Stress cardiomyopathy, also called apical ballooning syndrome, broken heart syndrome, takotsubo cardiomyopathy (TCM), and stressinduced cardiomyopa-

Abbreviations:

AMI	Acute myocardial infarction
TCM	Takotsubo cardiomyopathy
LV	Left ventricle
LVOT	Left ventricular outflow trac
ECG	Electrocardiogram
MR	Mitral regurgitation
ECG MR	Electrocardiogram Mitral regurgitation

thy, is an increasingly reported syndrome, generally characterized by transient systolic dysfunction of the apical and/or mid segments of the left ventricle (LV). It mimics acute myocardial infarction (AMI) but presents in the absence of obstructive coronary artery disease.^[1] Patients who are in shock should undergo urgent echocardiography to determine if left ventricular outflow tract (LVOT) obstruction is present.

In patients with hypotension and severe or moderate-severe LVOT obstruction, inotropic agents should Özet– Takotsubo kardiyomiyopatisi (TKM) apikal balonlaşma ile seyreden nadir bir akut kardiyomiyopati türüdür. Hipotansiyon ile seyreden hastalarda sol ventrikül çıkım yolu (SVÇY) darlığını saptamak amacıyla acil ekokardiyografi yapılmalıdır. Bu komplikasyon TKM'li tüm hastaların %10–25'inde tanımlanmıştır. Ayrıca TKM'li hastalarda SVÇY obstrüksiyonunun erken tanısı ve takibinin önemi bilinmediğinde bazı hastalar akut TKM'den kaybedilebilmektedir. Bu yazıda, kardiyojenik şok ile seyreden, hemodinamisi inotrop ajanlarla bozulan, fakat sıvı yüklemesi ve beta bloker tedavi ile düzelen TKM'li bir hasta sunuldu.

be avoided, because they can worsen degree of obstruction.

CASE REPORT

A 77-year-old female without known history of coronary artery disease was admitted to emergency services complaining of rapid onset of severe chest pain following an argument with her family. The character of the pain was retrosternal squeezing sensation radiating to the jaw and left shoulder. She had no history of traditional risk factors for coronary artery disease. Her electrocardiogram (ECG) showed ST-segment elevation in leads V1-6, II, III, and aVF, consistent with AMI (Figure 1). Due to severe hypotension (73/45 mmHg via intra-arterial measurement) dopamine infusion was initiated to treat cardiogenic shock. Noradrenalin infusion was added due to resistant hypoten-

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sion. Coronary angiogram revealed patent coronary arteries with minor irregularities and a thrombolysis in myocardial infarction flow grade of III. Transthoracic echocardiography showed hypokinesis of apical and midventricular walls and a left ventricular ejection fraction of 30%, consistent with TCM. Systolic anterior motion of the mitral valve and moderate mitral regurgitation (MR) were recorded (Video 1^{*}). Echo-derived peak and mean LVOT gradient were 137/61 mmHg (Figure 2). After obtaining a diagnosis of LVOT obstruction, inotropic agents were stopped and 500 cc bolus of 0.9% NaCland 5 mg bolus of intravenous metoprolol treatment were administered due to deterioration of hemodynamic status. Tachy-cardia and hypotension eventually decreased and oral metoprolol treatment was initiated. Subsequent course



was uneventful. A repeat transthoracic echocardiography after 3 days showed no LVOT gradient but mild apical hypokinesis and mild MR. (Figure 3). Control ECG taken a month later showed complete resolution of ST-segment elevation (Figure 4).

DISCUSSION

Stress cardiomyopathy was first described in Japan. The term "takotsubo" is taken from the Japanese term for an octopus trap, which has a shape similar to apical ballooning configuration of LV in systole in the typical form of this disorder. In the most commonly described typical type of stress cardiomyopathy, the contractile function of the mid and apical segments of the LV are depressed, and there is hyperkinesis of the basal walls, producing a balloon-like appearance of the distal ventricle with systole.^[2]

Although clinical presentation of TCM may be

identical to AMI, suspected pathogeneses differ greatly. The etiology of Takotsubo cardiomyopathy remains speculative. Proposed mechanisms include multivessel coronary artery spasm, impaired cardiac microvascular function, and endogenous catecholamine-induced myocardial stunning and microinfarction.

ECG often reveals ST elevation (often precordial) during the acute phase, followed by T-wave inversion, QT prolongation, and sometimes Q waves during the subacute phase. A few studies have shown minor ECG differences between patients with TCM and AMI. Cardiac markers are usually elevated. However, the levels tend to be lower and normalize sooner than in AMI patients. Angiography is required for diagnosis, as there is no accurate way to reliably distinguish TCM from AMI using ECG or cardiac markers.^[3]

Hypotension may develop in patients with TCM due to left ventricular systolic dysfunction, due to significant MR secondary to systolic anterior motion of the mitral valve, due to dynamic LVOT obstruction, or due to a combination of these factors. Alternately, hypotensive patients presenting with chest pain and ischemic electrocardiographic changes may, of course, have cardiogenic shock due to acute coronary syndrome. It is clinically important to differentiate the cause of hypotension in such patients because immediate management varies, depending on the underlying etiology.^[4,5]

Following diagnosis of stress cardiomyopathy, therapy is determined by the patient's overall clinical condition. Patients who are in shock should undergo urgent echocardiography to determine if LVOT obstruction is present. In contrast to hypotension due only to pump failure, hypotension associated with LVOT obstruction should not be treated with inotropic agents, because they can worsen the degree of obstruction. Recommended approach to patients with moderate-to-severe LVOT obstruction includes use of beta-blockers, which can improve hemodynamics by causing resolution of the obstruction. In addition, in the absence of significant pulmonary congestion, the patient should be fluid resuscitated.^[6,7]

In patients with LVOT obstruction and severe hypotension who either do not tolerate or do not adequately respond to beta-blockers, an alpha agonist may be added with caution and close monitoring. Phenylephrine is a pure alpha-adrenergic agonist that may reduce the gradient by increasing afterload, thereby improving overall hemodynamics. This treatment may be helpful to support blood pressure while a beta-blocker is administered to reduce inotropy. However, in the present case, a pure alpha agonist was not used, and the patient responded well to serum fluid and intravenous beta-blocker treatment.

Kawaji et al. showed that LVOT obstruction is a temporary complication and relief was confirmed by echocardiography or catheterization during follow-up in all patients. There were no significant differences in most baseline characteristics, including echocardiographic findings and medications, between TCM patients with or without LVOT obstruction. However, the prevalence of moderate to severe MR, congestive heart failure, and hypotension were significantly higher in patients with LVOT obstruction than in patients without. The cumulative 3-year incidence of all-cause death was not significantly different between the 2 groups, with no cardiac deaths in either group. However, the cumulative 3-year incidences of hospitalization for congestive heart failure and recurrent TCM were significantly higher in patients with LVOT obstruction^[8]

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*Supplementary video file associated with this article can be found in the online version of the journal.

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Keywords: Beta-blocker treatment; echocardiography; inotropic agents; outflow obstruction; Takotsubo cardiomyopathy.

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