Transient left ventricular apical ballooning syndrome: first series in Turkish patients

Geçici sol ventriküler apikal balonlaşma sendromu: Türk hastalarda ilk olgu serisi

Uğur Arslan, Yusuf Tavil, Adnan Abacı, Atiye Çengel

Department of Cardiology, School of Medicine, Gazi University, Ankara, Turkey

Left ventricular apical ballooning syndrome ("takotsubo" cardiomyopathy) (ABS), a novel cardiac syndrome, is characterized by transient apical ballooning leading to apical systolic dysfunction. This syndrome usually mimics acute coronary syndrome (ACS) because of the symptoms, electrocardiographic (ECG) changes and moderate cardiac enzyme release, however the myocardial area affected from ABS does not belong to a single coronary artery territory, coronary angiogram usually does not reveal a significant coronary artery disease and the myocardial dysfunction is almost always reversible.

Takotsubo cardiomyopathy was first described in Japan by Satoh et al (1) and Dote et al (2) as left ventricle affected from ABS was resembling an octopus trap (in Japanese named "takotsubo"). After that, many case reports were reported in Japan, but few in Western countries. Herein, we report 4 cases of transient ABS from Turkey.

The characteristics of these patients who developed ABS are shown in Table 1. All these patients were female and had a triggering event, physical or emotional stress before symptoms. The ECG changes and troponin T (TnT) positivity were mimicking ACS however three of them had normal coronary angiogram and the other patient had non-critical stenosis. Typical left ventriculography appearance of ABS of patient 1 is shown in Figure 1.

The ABS is an acute onset syndrome with chest symptoms, electrocardiographic changes, and minimal increase in myocardial enzyme levels in serum mimicking acute myocardial infarction leading to transient left ventricular apical wall motion abnormalities in patients without significant stenosis on coronary angiogram. All physicians should know the existence of this disease because thrombolytic therapy may be given by mistake to these patients with no occlusion in their coronary arteries.

The ABS, first thought to occur only in Japan, has now been demonstrated in many countries (3-6). To our knowledge, this is the first series of ABS described in Turkish patients.

This case series showed the importance of physical and emotional stress in the development of ABS. Three of these cases developed ABC after operation under general anesthesia and the other - after death of her husband. This disease is most commonly seen in elderly women. In one review (7) investigating 7 case series, 82-100% of the patients affected from ABS were women. All our four cases were women, which is also consistent with literature reports.

The pathophysiology of ABS is not still clear because ABS is a novel syndrome and is not seen commonly maybe because it is an underrecognized disease as the symptoms, ECG changes and moderately elevated cardiac enzymes make the physicians consider ACS. However, there are many postulations about the pathophysiology of this disease in the literature.

One of the important findings in ABS is that most of patients have increased catecholamine levels after a stressful event. Wittstein et al (8) have demonstrated supraphysiologic catecholamine levels in patients with transient left ventricular dysfunction after emotional stress. Initial plasma levels of catecholamines of this category of patients were markedly higher than in patients presented with acute myocardial infarction.

Classical myocardial ischemia and necrosis cannot explain the pathophysiology of this disease because the coronary angiographies performed immediately after this disease reveal no obstructive lesion in the coronary arteries and the myocardial area affected from this disease does not belong to a single coronary artery territory.

The most popular hypothesis in the development of ABS is myocardial stunning due to an epicardial coronary artery spasm. Microvascular spasm was also assumed to be the cause of this disease. There are several questions about these postulations. Why do all the patients with a stressful event not develop ABS? Why this disease is seen most commonly in elderly women? Why is there a geometrical distribution of this disease in the ventricle and the area affected does not belong to a single coronary artery perfusion territory?

Merli et al (9) have clarified these questions. Elderly women usually develop abnormal basal/mid-septal thickening known as sigmoid septum, which can produce high intraventricular gradients. In the presence of raised catecholamine levels due to

Address for Correspondence: Yusuf Tavil, MD, Yeşiltepe sitesi 7. blok, Daire: 45 Emek, Ankara, Turkey
Tel.: +90 312 202 56 47 Fax: +90 312 212 90 12 E-mail: yusuftavil@gmail.com

Table 1. Patient characteristics

Parameters	Patient 1	Patient 2	Patient 3	Patient 4
Age, years	63	66	70	71
Sex	Female	Female	Female	Female
Co-existing disease	Diabetes mellitus	Hypertension	--	Hypertension
Symptoms	Chest pain and dyspnea	Chest discomfort	Dyspnea	Dyspnea and chest pain
Triggering event	Meningioma operation	Laparoscopic cholecystectomy	Death of her husband	Abdominal hysterectomy
Electrocardiogram	ST segment elevation in precordial leads	ST segment elevation and T wave inversion in precordial leads	Non-specific T wave changes in the precordial leads	ST segment elevation in leads V1-V4
Troponin T, ng/ml	1.32	0.88	1.12	1.56
Complication	--	--	--	Acute heart failure
Coronary angiography	Normal	Normal	30% stenosis in LAD	Normal
Control echocardiography	Normal systolic functions one month later	Normal systolic functions with left ventricular hypertrophy after 40 days	Normal systolic functions with left ventricular hypertrophy after 30 days	Left ventricular hypertrophy with basal septal thickening without any systolic dysfunction after 20 days

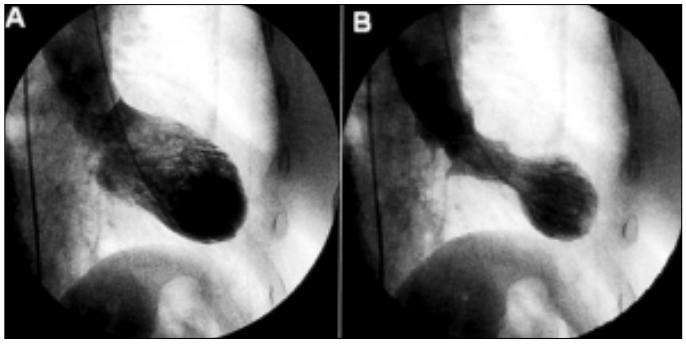

Figure 1. Typical appearance of apical ballooning seen in ventriculography. (A: Diastole B: Systole) The contraction of the basal segments of left ventricle is normal but dyskinesis is prominent in the apex making the apical segments of the left ventricle like a balloon

a stressful event, severe transient mid-ventricular obstruction, which divides the left ventricle into functionally different two cavities, apical chamber with high pressure and the proximal chamber with normal pressure, occurs in these patients. If dehydration (for example due to surgery) is also present, the gradients occurring in this obstructed area also increase contributing to increased pressure in the apical chamber. The combination of increased wall stress in the apical region and supraphysiologic catecholamine levels in plasma leads to subendocardial ischemia in the apex and myocardial stunning not related to a single coronary artery territory. When levels of catecholamines decrease, reversal of these events occurs and myocardial stunning due to ischemia resolves.

In our series, 3 women had left ventricular hypertrophy and 3 were after surgeries which favor this hypothesis. All these patients completely recovered echocardiographically after 3-5 weeks. Though we did not investigate the catecholamine levels, all of them had a stressful event in their history, which increases the catecholamine levels in plasma. Therefore, the importance of stress either physical or emotional should be emphasized in the pathophysiology of this disease. This syndrome can also explain deaths after stressful events because an ischemic substrate is present with very high catecholamine levels.

As a result, this is the first series of takotsubo syndrome described in Turkish patients. To know the existence of this syndrome is important for the clinicians especially in elderly female patients after a stressful event. Further studies are needed to explain the exact pathophysiology of transient left ventricular ABS.

References

1. Satoh H, Tateishi H, Uchida T, Dote K, Ishihara M. Takotsubo-type cardiomyopathy due to multivessel spasm. In: Kodama K, Haze K, Hon M, editors. Clinical Aspect of Myocardial Injury: from Ischemia to Heart Failure. Tokyo: Kagaku Hyoronsha; 1990. p. 56–64.
2. Dote K, Sato H, Tateishi H, Uchida T, Ishihara M. Myocardial stunning due to simultaneous multivessel spasms: a review of five cases. J Cardiol 1991; 21: 203–14.
3. Lupi G. Transient ballooning of the left ventricle: a case report. Ital Heart J 2004; 5: 635-7.
4. Kotzerke M, Keim M, Haller CH. Left ventricular apical ballooning. Dtsch Med Wochenschr 2004; 129: 2348-51.
5. Lupi G. Transient ballooning of the left ventricle: a case report. Ital Heart J 2004; 5: 635-7.
6. Seth P, Aurigemma G, Krasnow J, Tighe D, Untereker W, Meyer T. A syndrome of transient left ventricular apical wall motion abnormality in the absence of coronary disease: a perspective from the United States. Cardiology 2003; 100: 61-6.
7. Bybee KA, Kara T, Prasad A, Lerman A, Barsness GW, Wright RS, et al. Systematic review: transient left ventricular apical ballooning: A syndrome that mimics ST-segment elevation myocardial infarction. Ann Intern Med 2004; 141: 858-65.
8. Wittstein I, Thiemann D, Lima J, Baughman KL, Schulman SP, Gerstenblith G, et al. Neurohumoral features of myocardial stunning due to sudden emotional stress. N Engl J Med 2005; 352: 539-48.
9. Merli E, Sutcliffe S, Gori M, Sutherland GGR. Tako-Tsubo cardiomyopathy: New insights into the possible underlying pathophysiology. Eur J Echocardiogr 2006; 7: 53-61.