TAKO-TSUBO SYNDROME WITH DYNAMIC LEFT VENTRICULAR OUTFLOW TRACT OBSTRUCTION AND MITRAL REGURGITATION: A CASE REPORT

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Abstract

Tako-tsubo syndrome is characterized by acute chest pain, dynamic ST-T change and elevated cardiac markers, mimicking acute coronary syndrome. Severe, but transient LV regional dysfunction, more commonly in apical area, and patent coronary arteries establish the clinical diagnosis. The underlying mechanism of this syndrome is not fully clarified. Coronary spasm, microcirculation dysfunction and catecholamine overload have been proposed as the possible causes of this syndrome. LV outflow tract obstruction has been reported as a possible contributor to this syndrome. We report a case of tako-tsubo syndrome with markedly increased LV outflow tract pressure gradient and dynamic mitral regurgitation. The possible role of LV outflow tract obstruction in the pathophysiologic process is discussed. Echocardiography is a useful tool to identify this abnormality and to guide the therapy.

Key Words: Tako-tsubo cardiomyopathy, Apical ballooning syndrome, LVOT obstruction

Introduction

Tako-tsubo syndrome, also called apical ballooning syndrome, is characterized by acute regional left ventricular dysfunction (typically apical dyskinesia with compensatory basal segments hyperkinesia). It has a clinical presentation of acute chest pain with ST-T change on ECG mimicking an acute coronary syndrome. It is more common in elder women and is frequently precipitated by an emotional stress. The underlying mechanism of this syndrome is still not clear. We present here a case of tako-tsubo syndrome with high left ventricular (LV) outflow tract pressure gradient and discuss the possible contribution of LV outflow tract obstruction to the development of tako-tsubo syndrome.

Case Report

A 72 year-old woman presented to the emergency department with acute onset of chest tightness when she was working in the garden. The pain was compressive in character with radiation to her back and last for an hour. She denied any emotional stress at the time of symptom onset. A cardiac cath exam performed 3 years before this episode for her chest pain revealed patent coronary
arteries. An echocardiography at the same hospitalization showed good contractility, mild mitral regurgitation, without any evidence of LV outflow tract obstruction. On arrival, physical examination revealed a grade 2/6 pansystolic murmur over apex to left anterior axillary line and mild bilateral basal crackles. She had to keep two pillows for orthopnea. The serum level of troponin I on arrival was elevated at the level of 4 ng/ml. A twelve-lead ECG showed sinus rhythm and borderline ST elevation over lead II and aVF. Mild congestion over bilateral lung field was found in a chest X-ray exam. An echocardiography revealed apical dyskinesia, basal hyperkinesia, and a severe mitral regurgitation. An intra-ventricular pressure gradient up to 63mmHg at LV outflow tract over the bulging septum, and the systolic anterior movement of mitral leaflets highly suggested the outflow tract obstruction (Fig. 1). An emergent coronary angiography confirmed the patent coronary arteries. Right heart catheterization revealed mean pulmonary artery wedge pressure of 14mmHg and a giant v wave, compatible with the severe mitral regurgitation. The symptoms improved in the first few hours after admission. A tranesophageal echocardiography was performed on the second day of hospitalization but only mild mitral regurgitation was found. The 12-lead ECG on the second day showed new deep T wave inversion over V2-6 (Fig. 2). Carvedilol was administrated for the outflow tract obstruction. A follow-up echocardiography demonstrated no detectable intra-ventricular pressure gradient but persistent apical dyskinesia one week after admission.

Discussion

The mechanism of Tako-Tsubo cardiomyopathy is not fully understood. The prevalence of this syndrome is low. In the patients presented with symptoms suggestive of acute coronary syndrome, only 0.7 - 2.5% was diagnosed as tako-tsubo syndrome. Because of the low prevalence rate, there is still no enough data to confirm the underlying causes. There are several possible mechanism been proposed, including coronary artery spasm,
microcirculation dysfunction and catecholamine overload.¹ There are also four reports including 25 cases in the literature till now showing that LV outflow tract obstruction might be present in the tako-tsubo syndrome and might contribute to the pathogenesis of this syndrome. It is proposed that an increased LV outflow tract pressure gradient imposes a great wall stress to the apical area. This increased wall stress may results in an inadequate subendocardial coronary flow in the apical area. Villareal et al. first reported 3 patients with LV outflow tract obstruction in tako-tsubo syndrome presented with chest pain after emotional stress. Suppression of contractility with beta-blockers resulted in the improvement of LV outflow tract obstruction and in clinical symptoms.² Merli et al. described 4 women of typical tako-tsubo syndrome with a mid-cavity dynamic obstruction. The dynamic obstruction, which might be related to the localized mid-ventricular septal thickening, resolved prior to the resolution of the LV wall motion abnormalities. After improvement in wall motion, they performed a low-dose dobutamine stress echocardiography to reproduce the LV mid-cavity pressure gradient. This stress test showed the typical apical regional motion abnormalities in acute presentation. They suggested that the presence of a localized septal hypertrophy might be an important factor in the development of tako-tsubo syndrome.³ In the retrospective observational study, Mahmoud et al. reported 8 women of tako-tsubo syndrome with LV outflow tract obstruction. The prevalence of LV outflow tract obstruction in their series was 25% (8 in 32). Patients of tako-tsubo syndrome with LV outflow tract obstruction were characterized by older age, high New York Heart Association functional class, higher rate of septal bulge and higher degree of mitral regurgitation.⁴ In the initial report from Japan, there were 13 out of 72 patients with documented transient intraventricular pressure gradient.⁵ In spite of these report and hypothesis, the casual relationship between LV outflow tract obstruction and the development of tako-tsubo syndrome is still not established. It is possible that tako-tsubo cardiomyopathy is not a homogenous disease but a syndrome with different pathophysiologic processes. However, judging from the high level of LV outflow tract pressure gradient and it dynamic feature, it is highly possible that this obstruction contribute partly to the development of regional dysfunction. Besides, inotropics should be avoided in this syndrome if associated with an increased LV outflow tract pressure gradient.

Our patient has the typical characteristics

Fig. 2. T wave inversion over V2-V6.
LVOT obstruction in tako-tsubo cardiomyopathy

of the tako-tsubo cardiomyopathy. However, no emotion stress before this attack was reported by our patient. The LV outflow tract obstruction was dynamic, and the pressure gradient recorded when performing echocardiography was probably less than the peak level when symptoms were most severe. The dynamic nature of the mitral regurgitation was also confirmed from serial echocardiography examination. Mitral regurgitation associated with the LV outflow tract obstruction is probably related to the anterior approximation of mitral leaflets, which leads to mal-coaptation of mitral leaflets. The decreased severity of mitral regurgitation was parallel to the improvement in the symptom in our patient. Beta-blocker was given early after the finding of LV outflow obstruction. The resolution of LV outflow tract obstruction before that of apical dyskinesia in this patient also suggested that obstruction may contribute to the development of apical dyskinesia.

In conclusion, we suggest the early and repeated echocardiography exam in patients with tako-tsubo cardiomyopathy and a careful search for the LV outflow tract pressure gradient, especially when associated with a bulge septum and mitral regurgitation. Beta-blockade may be helpful in this situation. Further studies are needed to clarify the role of LV outflow tract in the development of tako-tsubo syndrome.

References

章魚壺心肌症合併左心室出口阻塞及二尖瓣逆流 — 病例報告

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摘要

章魚壺心肌症臨床表現類似急性冠心症，包括急性胸痛，ST-T 節段動態變化及心肌損傷指標的上升。若合併局部心臓收縮功能不良（特別在左心室心尖）及正常的冠狀動脈則可診斷此疾病。章魚壺心肌症的原因及病理生理機轉目前仍未完全明瞭。目前有幾種學說被提出，包括冠狀動脈痙攣、冠狀動脈微循環功能異常及血中兒茶酚胺激素過高，用來解釋此一症候。左心室出口壓力增加也在章魚壺心肌症的病患中被觀察到，也被提出用來解釋其病理生理機轉。我們報告一例章魚壺心肌症的病患合併有明顯增高的左心室出口壓力及動態的二尖瓣逆流，並討論左心室出口壓力增高在此症候群可能扮演的角色。心臓超音波檢查在此類病患為重要的診斷工具，並可能影響治療方向。

關鍵詞：章魚壺心肌症，左心室出口阻塞

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