

# A Unique Case Of Recurrent Takotsubo Cardiomyopathy- Atypical Followed By Typical Variant.

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## Abstract

Takotsubo cardiomyopathy (TC), synonymous with apical ballooning syndrome, broken heart syndrome, stress induced/ ampulla cardiomyopathy is characterized by transient left ventricular dysfunction of apical ( typical) or mid segments (atypical), mimicking acute myocardial infarction in the absence of significant coronary artery disease . We report a rare case of recurrent takotsubo cardiomyopathy in which the first and second presentations were atypical and typical variants respectively. We performed a literature search regarding takotsubo cardiomyopathy with emphasis on incidence and recurrence especially if one type was followed by other.

## INTRODUCTION

Since the first case reported by Sato et al [1] about two decades ago, takotsubo cardiomyopathy has gained significant interest for its presentation mimicking acute coronary syndrome without significant obstructive coronary artery disease and at the same time having remarkable wall motion abnormalities which resolves in the ensuing few weeks. The condition is also known as broken heart syndrome, ampulla cardiomyopathy and stress induced cardiomyopathy [2,3].

## CASE REPORT

A 51 year old female with past medical history of ulcerative colitis s/p colectomy and ileostomy and no history of cardiac disease presented to the hospital with chief complaint of abdominal pain. A CT scan of the abdomen was done which revealed a calculus at the right uretero-vesical junction with moderate right hydronephrosis. The next day of admission, the patient was to be taken to the operative room for a ureteral stent placement but while waiting for the same she complained of sudden onset of shortness of breath. Vital signs at that time were : BP 130/70 heart rate 130/min, RR 22/min, O2 saturation - 100% on 4 litres of oxygen. An electrocardiogram (ECG) showed ST depression in leads V3-V6 . Chest X-ray revealed pulmonary edema. Laboratory values were as follows: WBC- 19,900/mm<sup>3</sup>, Hgb -12.6 mg/dL, Na- 139 mEq/mL, K- 3.5 mEq/mL, HCO<sub>3</sub>- 17 mEq/mL, BUN 17 mg/dL, creatinine 1.7 mg/dL. Troponin I was found to be elevated at 14.7 ng/ml (normal <0.1). The

patient was given i/v furosemide which alleviated her respiratory distress and she was taken for cardiac catheterization which revealed normal coronaries , and left ventriculogram showed severe basal akinesis, anterior and lateral akinesis but normal apical motion (Figure 1A and 1B) (atypical variant of TC). The patient was treated conservatively with diuretics and beta-blocker and she recovered without further complications. Over the next few days ECG reverted back to normal and a follow up echocardiogram done 2 weeks later showed that wall motion abnormalities had disappeared and left ventricular ejection fraction (LVEF) which was estimated at 30% during the acute event had normalized to 60%.

Exactly three years later, patient presented with nausea, vomiting and abdominal pain. A CT scan for abdomen suggested ileus and bilateral renal calculi which were non-obstructive. She was admitted for intra-venous hydration and pain management . While on the floor, she had sudden onset of left sided chest pain and shortness of breath. Vital signs were as follows- BP 114/65 mm Hg, HR 124/min, RR 22/min, O2 saturation 92% on 100% non-rebreather mask. An ECG revealed ST elevation in leads V3-V6. Laboratory values were as follows- WBC 5,200/mm<sup>3</sup>, Hgb 12.4 mg/dL, BUN 9 mg/dl, creatinine 0.9 mg/dL and troponin I was elevated at 6.1 ng/ml. Patient was immediately taken for cardiac catheterization which showed normal coronary arteries and left ventriculogram revealed hypercontractile basilar segments and the rest of the left ventricle was akinetic

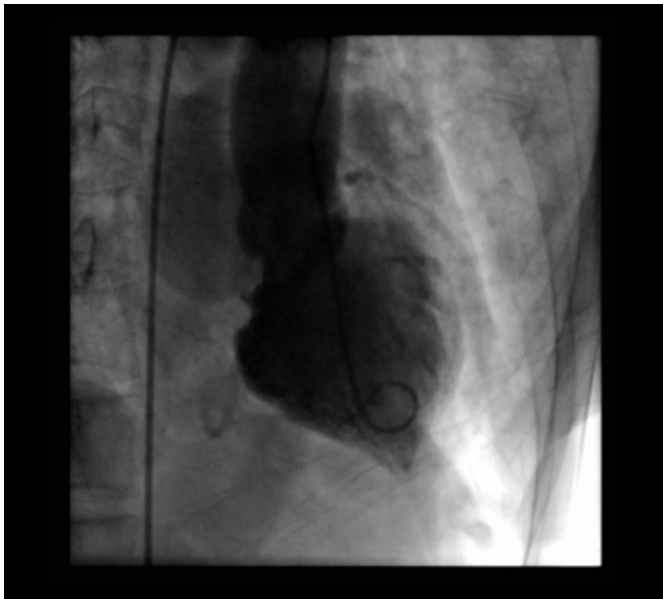
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(Figure 2A and 2B) (typical variant). LVEF during this admission was found to be severely depressed at 10-15%. She was treated conservatively with diuretics and beta-blocker again. The patient recovered fully, repeat ECG over the next few days showed resolution of ST elevation and a follow up echocardiogram done a week later showed resolution of wall motion abnormalities and LVEF was estimated at 55-60%. The patient was discharged after resolution of ileus without any further untoward incidents.

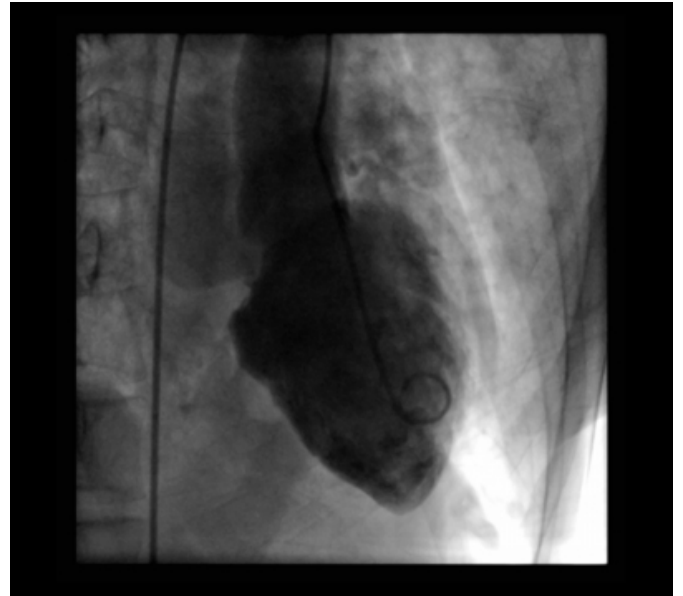
**Figure 1**

Figure 1A – Right Anterior Oblique (RAO) end systolic left ventriculogram in atypical variant of TC.



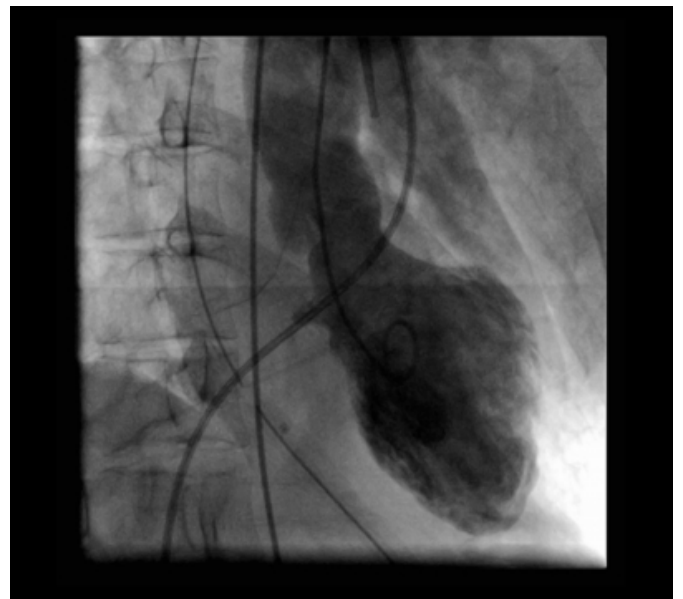
**Figure 2**

Figure 1B - RAO end diastolic ventriculogram in Atypical variant of TC.



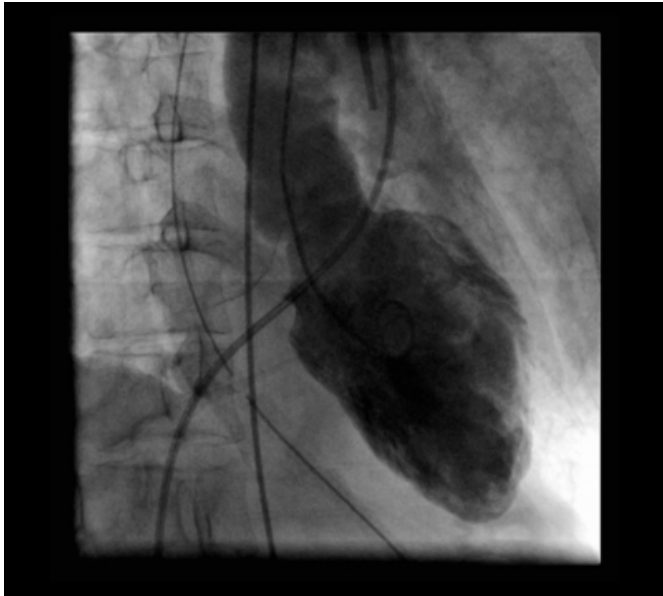
**Figure 3**

Figure 2A- RAO end systolic left ventriculogram in typical variant (basal ballooning) of TC.



**Figure 4**

Figure 2B– RAO end diastolic ventriculogram in typical variant of TC.



**DISCUSSION**

Takotsubo cardiomyopathy (TC) derives its name from Japanese octopus trap, which has a shape similar to apical ballooning configuration of the left ventricular systole in the ‘typical’ form of this disorder. In a minority of cases (40% in one report) [4], defined as atypical, ventricular hypokinesis is restricted to mid ventricle with relative sparing of the apex. A prevalence of 1.2% was reported from a registry of 3265 patients with troponin positive acute coronary syndrome[4]. A recent study conducted by Elseber et al reported a recurrence rate of 11.4% over 4 years after initial presentation, with an average recurrence rate of 2.9% over first few years and subsequently decreased to 1.3% per year[5]. TC is unique in that it usually occurs in post-menopausal women with more than 90% of reported cases being their population (6,7). The mean age has been ranged between 58-75 years with just about 3% patients being less than 50 years(7). Interestingly younger premenopausal females present with atypical variants (8). Time to recurrence has been variable from 3 months to 13 years, in our case it was 3 years. The onset is frequently triggered by an acute medical illness or by intense emotional or physical stress. Pathogenesis of this disorder is not well understood and postulated mechanisms include catecholamine excess, multiple coronary artery spasms and microvascular dysfunction resulting in myocardial stunning[7]. No obvious mechanism for the different patterns of regional wall motion abnormality i.e for typical and atypical patterns has been identified.

ECG abnormalities are the most common finding. In a systematic review ST elevation was present in 56 % ( our patient had this finding during second presentation with typical pattern) and among these it was most common in the anterior precordial leads[8]. The remaining patients had deep T wave inversion, abnormal Q waves, non-specific abnormalities and normal ECG on presentation[10]. Echocardiography or left ventriculography usually shows the characteristic apical ballooning with akinesis or dyskinesis of the apical one-half to two-thirds of the LV with an average EF ranging from 20-49% ( our patient had 10-15% and 30% on the two occasions). Emerging evidence shows involvement of right ventricle as well and its involvement has a negative impact on hospital stay and morbidity, and its identification can help predict hemodynamic instability [11]. Troponin elevation is usually modest and hemodynamic compromise is out of proportion.

Suspicion of diagnosis of TC is not a sufficient reason to withhold PCI or fibrinolytic therapy since the majority of ST elevation presentation will be due to a critical coronary lesion. Those presenting without ST elevation will usually fit into a high risk NSTEMI profile and early cardiac catheterization will be performed.

Despite the severity of the acute illness, takotsubo cardiomyopathy is a transient disorder managed with supportive therapy. Resolution of the physical or emotional stress usually results in rapid resolution of symptoms and ECG changes. Patients are usually treated with standard heart failure medical regimen inclusive of diuretics, angiotensin converting enzyme inhibitors and beta-blockers[12] until there is recovery of systolic function which occurs in 1-4 weeks in most cases. Because the condition may recur, adrenergic blockade is usually continued indefinitely. Our patient was not on adrenergic blocker on second presentation, however no difference was observed between patients with and without recurrence in the use of beta-blockade, ACE-I and statins.[5].

Our patient had a unique presentation of atypical and typical wall motion abnormalities on first and second occasions respectively, it is very difficult to explain the mechanism as the precipitating factor i.e abdominal pain appeared to be the same. ECG changes were seen in the same leads i.e V3-V6 but was ST depression in the first event and ST elevation followed by T wave inversion before reverting back to normal during the second occurrence. Kurisu et al stated that admission ECG showed ST elevation or T wave inversion in V3-V6 and then T wave became inverted within 2 days and

then progressed to a negative peak in about 3 days [13] which was seen in our patient. One thought is that first excessive catecholamine stimulation lead to down-regulation of receptors of the affected area in the first instance and the reactivity or sensitivity difference to the second excessive catecholamine stimulation might present an opposite picture[14]. Hurst et al reported that the difference in wall motion abnormalities of LV apical ballooning and mid-ventricular ballooning reflect temporal variation in the resolution of apical ballooning at the time of angiographic diagnosis[15].

Defining the mechanism of takotsubo cardiomyopathy and may be different for typical and atypical variants is necessary to prevent recurrences. How frequently the two variants follow each other in recurrence is not known? As of yet we feel that this is very rare and the first case of atypical being followed by typical variant as per our literature review. On the basis of this case report, it does not seem that distribution of cardiac sympathetic nerves is a mechanism to explain the enigma called 'takotsubo cardiomyopathy'.

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