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Apical Ballooning Syndrome: A Cardiac Syndrome Mimicking Acute Myocardial Infarction

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Abstract **Case Report**

Introduction: Apical ballooning syndrome, Takotsubo cardiomyopathy, or broken heart syndrome, is a rare but currently well recognized clinical entity. It occurs after severe emotional or physical stress, most often in postmenopausal women, and mimics the symptoms and signs of acute coronary syndrome, in the absence of significant coronary injury. This cardiomyopathy is rapidly and completely reversible. The exact pathophysiological mechanism of the condition remains debated; several hypotheses have been put forward, the most credible of which seems to be a sudden hyperadrenergic reaction related to stress. ***Objective:*** To report the case of a patient with clinical features of possible myocardial infarction but diagnosed as Takotsubo cardiomyopathy. ***Case Report:*** The patient was a 64-year-old woman with no cardiovascular risk factors other than age and menopause, and no particular pathological history. She was admitted on Day 3 with infarct-like chest pain, which appeared following an episode of emotional stress, associated with NYHA stage III dyspnea. On admission, the patient was pain free. The clinical examination found a conscious patient, hemodynamically and respiratory stable, apyretic. The cardiovascular examination was unremarkable. The ECG was in sinus rhythm, showing a QS aspect in the anterosepto-apical area associated with negative T waves in the inferolateral area. The biological workup showed an inflammatory syndrome, with positive troponin at 650ng/l. The thoracic echocardiography showed an aspect of hypokinetic cardiopathy at the dilated stage with severe LV dysfunction (LVEF at 38%), a thin and flexible mitral valve with restriction of the small mitral valve with moderate mitral insufficiency (SOR at 20 cm2 and RV at 19 ml), a dilated left atrium. Coronary angiography showed angiographically healthy coronary arteries, and an MRI was performed, confirming the diagnosis of Takutsbo cardiomyopathy. ***Discussion and Conclusion***: Takotsubo cardiomyopathy is an increasingly recognized clinical entity, characterized by apical left ventricular dysfunction with chest pain, electrocardiographic changes, minimal biomarker changes, and no coronary lesions. In most cases, an episode of emotional or physical stress precedes the event. Its pathophysiological mechanism remains unknown, but high circulating catecholamines seem to be the triggering factor and the most plausible explanation for this primary acquired cardiomyopathy. It is important to be aware of this diagnosis in patients presenting with ACS because its evolution is most often spontaneously favorable.

Keywords: Takotsubo cardiomyopathy, acute coronary syndrome, coronary angiography, prognosis.

INTRODUCTION

Apical ballooning syndrome, Takotsubo cardiomyopathy, or broken heart syndrome, is a rare but currently well recognized clinical entity [1]. It occurs after severe emotional or physical stress, most often in postmenopausal women, and mimics the symptoms and signs of acute coronary syndrome, in the absence of significant coronary injury. This cardiomyopathy is rapidly and completely reversible. The exact pathophysiological mechanism of the condition remains debated; several hypotheses have been put forward, the most credible of which seems to be a sudden hyperadrenergic reaction related to stress [2].

We report the case of a 64-year-old female patient with clinical features of possible myocardial infarction but diagnosed as Takotsubo cardiomyopathy.

CASE REPORT

The patient was a 64-year-old woman with no cardiovascular risk factors other than age and menopause, and no particular pathological history. She was admitted on Day 3 with infarct-like chest pain,

which appeared following an episode of emotional stress, associated with NYHA stage III dyspnea. On admission, the patient was pain free.

The clinical examination found a conscious patient, hemodynamically and respiratory stable, apyretic. The cardiovascular examination was unremarkable.

The ECG was in sinus rhythm, showing a QS aspect in the antero-septal area associated with negative T waves in the inferolateral area (figure 1).

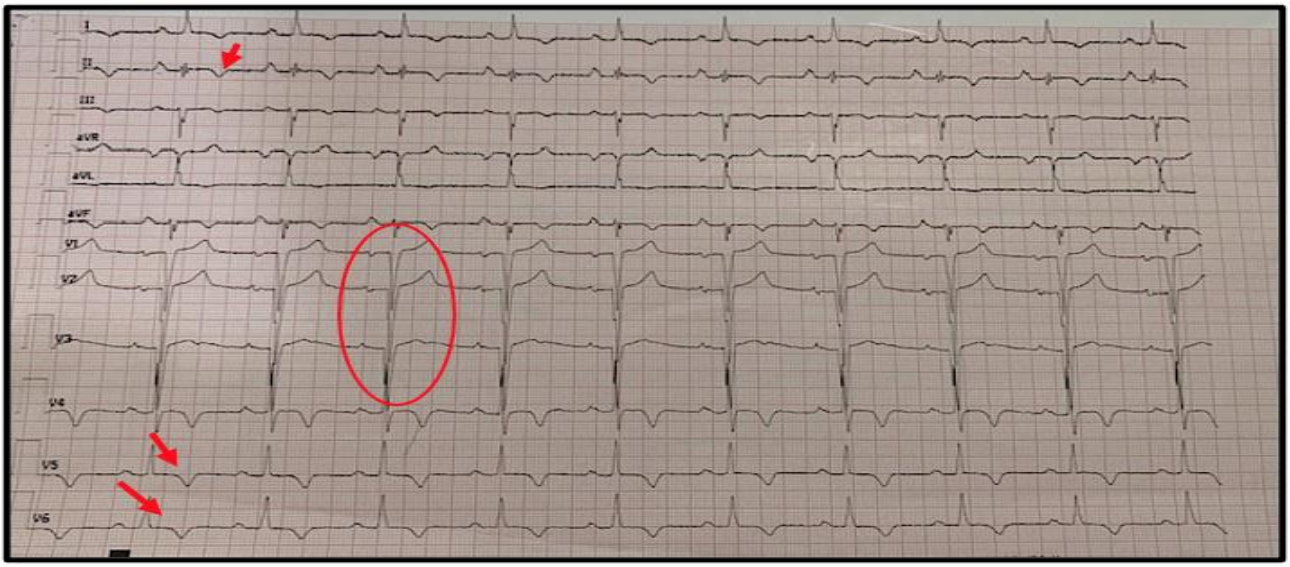

Figure 1: ECG showed an anteroseptal QS appearance associated with negative T wave's infero-lateral

The biological workup showed an inflammatory syndrome, with positive troponin at 650ng/l.

The thoracic echocardiography showed an aspect of hypokinetic cardiopathy at the dilated stage with severe LV dysfunction (LVEF at 38%), a thin and flexible mitral valve with restriction of the small mitral valve with moderate mitral insufficiency (SOR at 20 cm2 and RV at 19 ml), a dilated left atrium (figure 2).

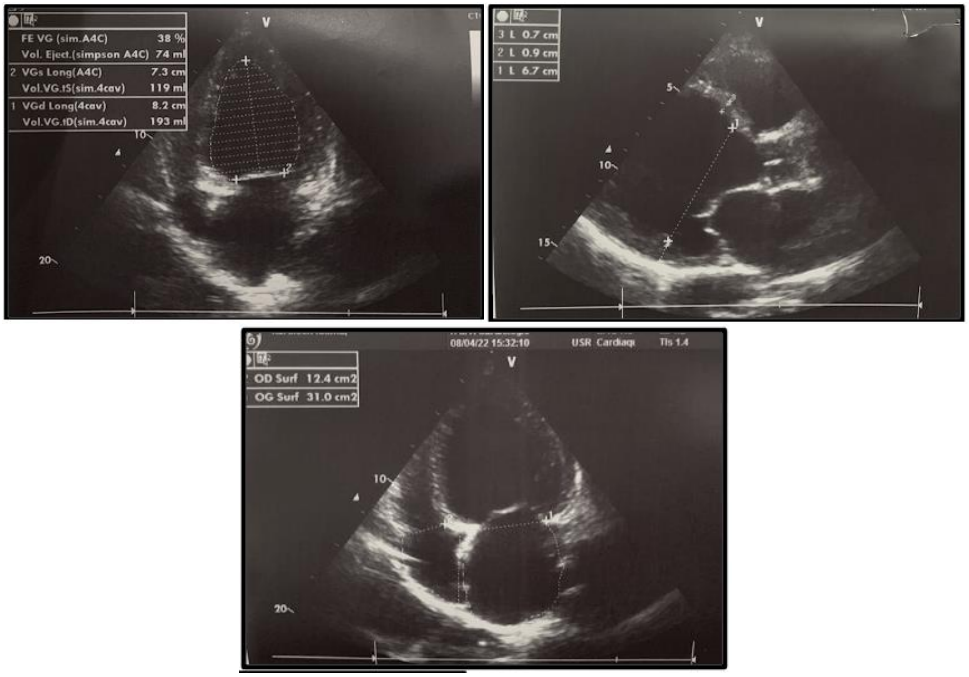

Figure 2: TTE showed hypokinetic heart disease at the dilated stage with severe LV dysfunction (LVEF 38%)

Coronary angiography showed angiographically healthy coronary arteries (Figure 3).

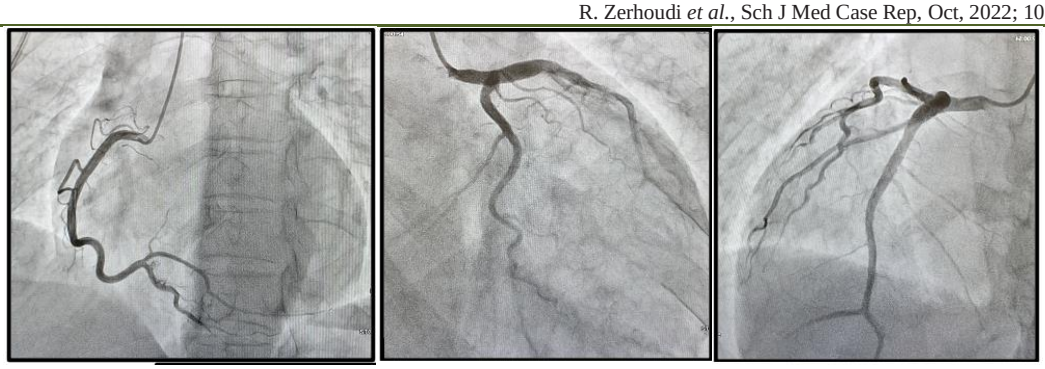

Figure 3: Conary angiography performed in the emergency setting showed angiographically healthy coronary arteries

The evolution was marked by a clinical improvement under heart failure treatment, an echocardiography performed after 6 months showed an improvement of his LVEF (figure 4).

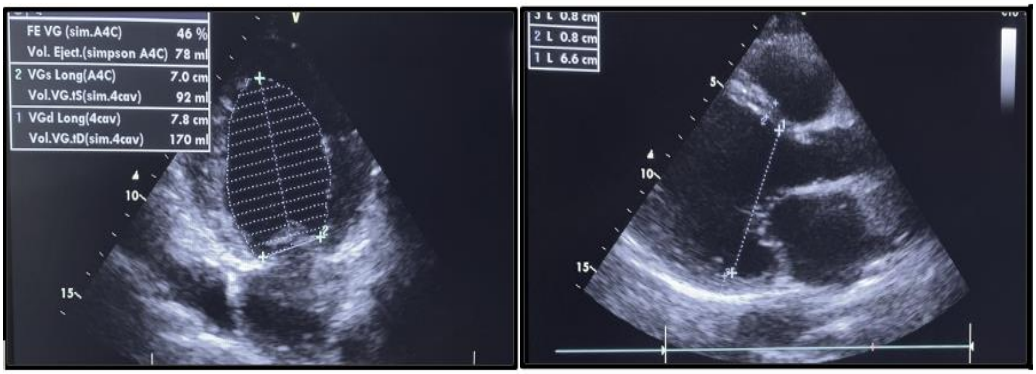

Figure 4: Control TTE showed improvement in LVEF (LVEF at 46%)

An MRI was performed, confirming the diagnosis of Takutsbo cardiomyopathy.

DISCUSSION

Takotsubo cardiomyopathy was described in 1990 by Sato [2]. The first cases described in Belgium were reported in 2003 by Desmet [3]. The choice of the name "Takotsubo" comes from the resemblance of the left ventricle, objectified during ventricular angiography, with the shape of the vases used as octopus traps by Japanese fishermen (in Japanese, octopus is called "tako" and vase is called "tako")(in Japanese, octopus is called "tako" and vase is called "tsubo"). Its exact prevalence is unknown but it is thought to affect 1 to 2% of acute coronary syndromes [4-6].

The literature essentially reports isolated clinical cases or limited series of patients (< 15 patients); the only exception is a Japanese review, the most important, which includes 88 patients [6]. It should be noted that Gianni analyzed the fourteen most relevant studies among all the literature [3].

Tako-tsubo cardiomyopathy presents as an acute coronary syndrome with acute constrictive resting chest pain, most often occurring after significant emotional or physiologic stress [3, 7]. The majority of patients are postmenopausal women, between 62 and 75 years of age, although extremes of 10 to 88 years have been reported [3, 4, 6]. Sometimes, the initial clinical picture reflects severe complications, such as cardiogenic cardiogenic shock. This prevalence, from 2% to 46%, varies greatly from one series to another [4, 6]. The occurrence of ventricular arrhythmias is estimated at 1.5% (from 0.65% to 3.9%) [4]. A case of QT interval prolongation and torsades de pointes were described in 2008 (8). In the Tsuchihashi series, the in-hospital mortality rate is estimated at 1% [6].

Thus, the authors of the Mayo Clinic proposed 4 criteria necessary for diagnosis [6]:

1. Transient akinesia or dyskinesia of the apical and middle segments of the left ventricle whose extent does not correspond to a coronary territory.
2. Absence of stenosing coronary artery disease or angiographic evidence of acute plaque rupture.
3. Appearance of electrocardiographic abnormalities (either ST-segment elevation or T-wave inversion)
4. Absence of:

- Recent head trauma
- Intracerebral hemorrhage
- Pheochromocytoma
- Myocarditis
- Hypertrophic cardiomyopathy

Initially, Takotsubo cardiomyopathy was considered to specifically affect the apical 2/3 of the left ventricle (apical ballooning of the LV) (Figure 5). Then

came the basal forms called "reverse takotsubo" or "squidd syndrome", a name given by Japanese authors, by analogy with the squid form (figure 6).

Medioventricular forms were then described, with circumferential akinesia of the middle part of the left ventricle and hyperkinesia of the apex and base (Figure 7). It seems that the basal forms are rather the prerogative of younger patients [9].

In fact, only the circumferential character seems to be a common and characteristic criterion of Takotsubo cardiomyopathies. In general, it is easy to differentiate these pathologies from coronary territory involvement.

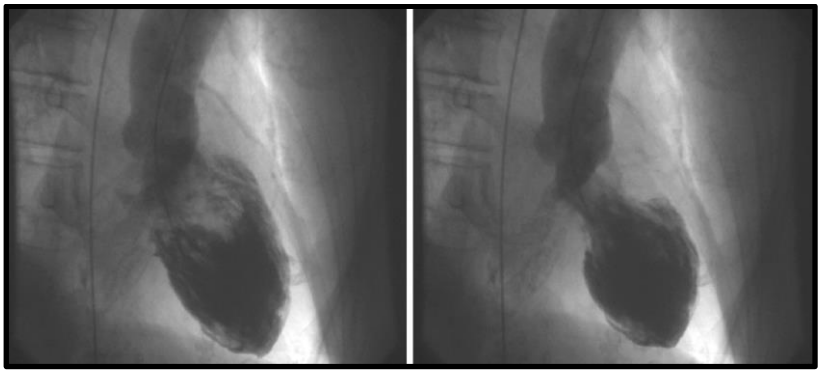

Figure 5: Typical appearance in diastole and systole of Takotsubo of the apical 2/3 "apical ballooning syndrome"

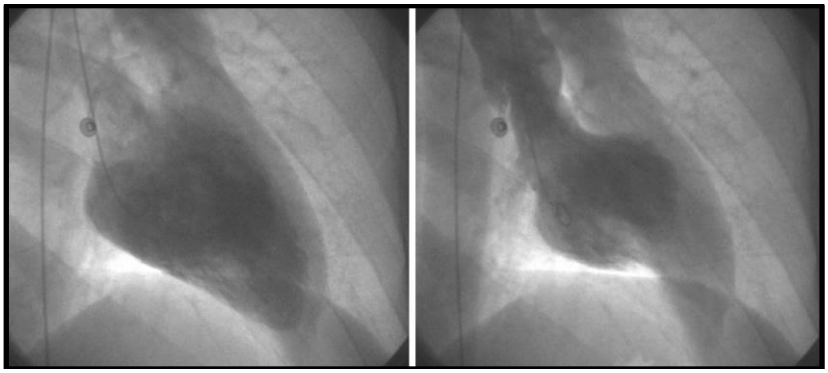

Figure 6: Typical diastolic and systolic appearance of a 2/3 basal Takotsubo ("squidd syndrome")

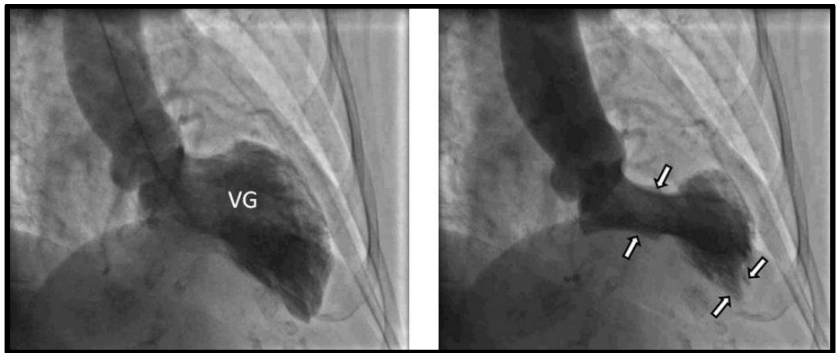

Figure 7: Midventricular forms with circumferential akinesia of the middle part of the left ventricle and hyperkinesia of the apex and base

The clinical presentation of Takotsubo cardiomyopathy is usually that of an acute coronary syndrome [10]. Thus, the majority of patients have angina-like chest pain. Other clinical signs are dyspnea (a sign of heart failure), and more rarely syncope (in case of rhythm disorder) or sudden death.

A stress triggering cardiomyopathy is found in the vast majority of cases (in more than 80%). It can be an emotional stress, such as an aggression, the announcement of a death of a close relative or a physical stress such as a fire, a road accident. Takotsubo cardiomyopathy can also occur after an acute illness (asthma, epileptic seizure) or a medical-surgical procedure (invasive, semi-invasive examinations and surgery). This stress can sometimes be minimal, and even the succession of seemingly anecdotal stresses has been found. However, the presence of stress is not systematic [11].

Various ECG abnormalities can be observed. None of them is specific. The most frequent are ST-segment elevation, most often anterior but sometimes more diffuse. The evolution is often made in a few hours towards the appearance of large negative T waves in the anterior part. A prolongation of the QT space is frequent. Repolarization disorders may persist for several weeks.

Markers of necrosis (troponin, CPK) are moderately increased. CPK levels are usually below 600. Normal CPK or troponin levels can be found even in cases of severe angiographic damage and therefore do not rule out the diagnosis. BNP elevation is only a consequence of hemodynamic alteration but may allow monitoring of the proper course in case of hemodynamic failure. Epinephrine, norepinephrine, and dopamine levels are elevated during the first 48 hours but are not routinely measured.

Transthoracic echocardiography is also a valuable aid, especially in cardiac intensive care and in the ICU, to assess left ventricular systolic function (Figure 8) and to search for a possible complication. The use of new techniques such as longitudinal myocardial deformation tools (strain) and contrast agents allowing opacification of the left heart chambers may improve the performance of traditional echocardiography (Figure 9) [12-14].

Finally, echocardiography allows monitoring of the left ventricular ejection fraction and thus ensures recovery ad integrum of this cardiomyopathy.

At the acute stage, typical abnormalities of segmental kinetics are demonstrated: a large apical akinetic zone with hyperkinesis of the basal segments. The left ventricular ejection fraction is decreased, with a mean of 20-40% [6]. Most patients recover normal function within days to weeks. Echocardiography can also be used to identify certain complications (mitral insufficiency, pericardial effusion, etc.) and serves as a non-invasive reference for subsequent follow-up.

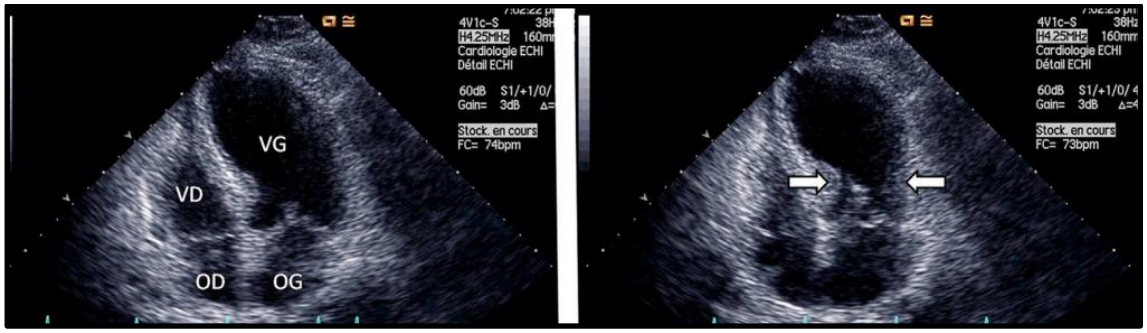

Figure 8: Apical four-chamber section in diastole (A) and systole (B) showing akinesia of the different middle and apical segments of the left ventricle (LV), with contractility only of the basal collar (arrows)

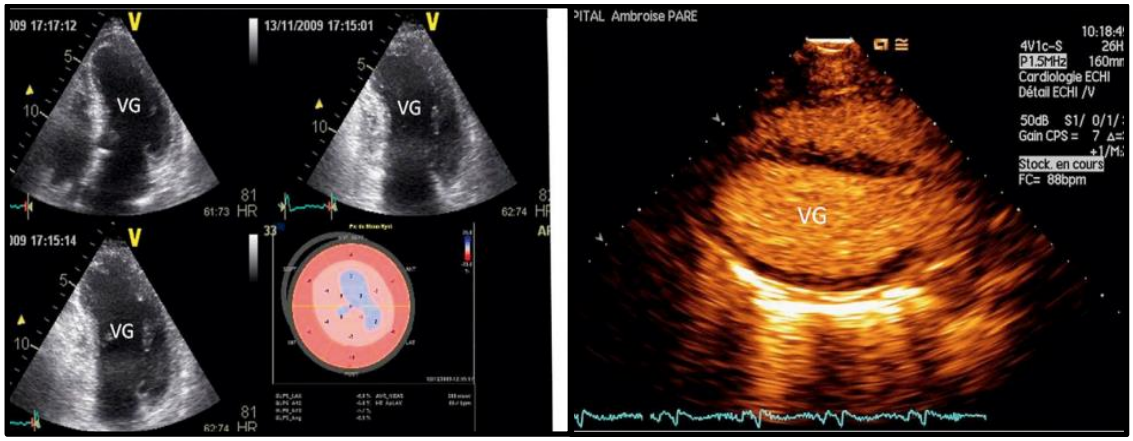

Figure 9: Myocardial deformation tool (strain) to demonstrate circular impairment of left ventricular dysfunction. D. Use of specific contrast agent (opacification of the left heart chambers) allowing better visualization of left ventricular dysfunction

MRI is frequently used for diagnosis, and many papers have been published on this examination in Takotsubo cardiomyopathy. It shows the geography of left and right ventricular involvement, as well as evidence of myocardial edema (Figure 10). The absence of late gadolinium enhancement in Takotsubo cardiomyopathy, unlike myocarditis, remains controversial.

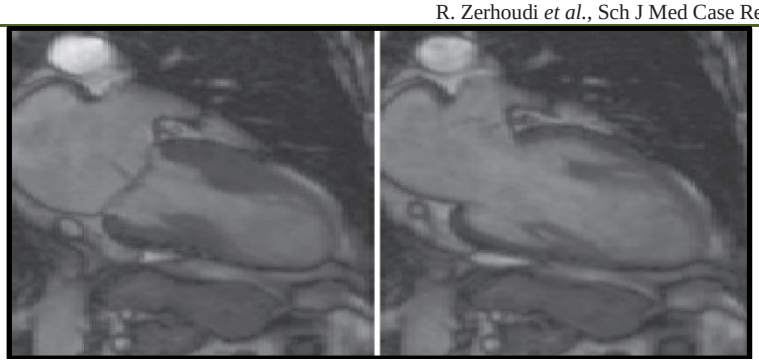

Figure 10: Appearance of Takotsubo on cardiac MRI

It seems difficult to affirm with certainty the diagnosis of Takotsubo cardiomyopathy without coronary angiography, not to rule out coronary disease, but to prove the discordance between the kinetic disorder and the coronary anatomy. One may be struck by the frozen appearance of the middle and apical portion of the coronaries, contrasting with the hypermobility of the base, especially when the right coronary is opacified first, in OAD, in a context of suspected anterior infarction.

Left ventricular angiography has the advantage of being very simple, inexpensive, and can be performed at the same time when the diagnosis is suspected during emergency coronary angiography. It should be performed in biplane, with the incidence in OAD, in cases of spasm of an IVA largely bypassing the tip, which may ape Takotsubo cardiomyopathy. Right ventricular angiography is more difficult to evaluate but is abnormal in at least 1/3 of cases.

Différentes explications physiopathologiques ont été évoquées. L'hypothèse de spasmes épicardiques multiples évoqué dans les premières publications et le rôle de l'obstruction la chambre de chasse du VG ne semblent guère plausibles.

Direct myocardial toxicity of catecholamines has been suggested by histological studies but may play only a secondary role. The role of a sudden and very high blood pressure surge (how high can the blood pressure be raised?) has never been mentioned.

A dysfunction of the myocardial micro-vascularization has been suspected and highlighted by some authors [15]. It would cause ischemia-reperfusion phenomena, a mechanism that would be roughly similar to the lesions observed in frostbite of the extremities, a pathology that is also "circumferential". It is the activation of the sympathetic nervous system that seems to play the primary role [16]. Experimentally, stimulation of the stellate ganglion causes pictures similar to Takotsubo cardiomyopathy and can be prevented by surgical (section of the ganglion) or pharmacological (propranolol, reserpine) methods [17].

The topography of Takotsubo cardiomyopathy could be explained by the local release of norepinephrine with a distribution related to nerve branches. It is possible that adrenaline released from the adrenal gland plays a triggering factor for a breakdown of the sympathetic system, making the humoral and neurogenic mechanisms intertwined.

The final mechanism is the same whether the stress is a single acute event or the result of minimal stress with a triggering threshold reached.

At the tissue level, histological analysis shows that myocardial cell architecture is preserved. The predominance in postmenopausal women would be related to the decrease in estrogens, which increases the action of hormones such as epinephrine on the heart. The predominance of involvement of the left ventricular apex is probably due to a difference in geographic response to catecholamines. Beta-receptor expression appears to be increased in the apical area, particularly in postmenopausal women.

A large number of complications have been described but most of them are not specific to the pathology. They are generally early and favored by the fact that we are dealing with a rather old population that suddenly presents a left ventricular dysfunction.

The first complication is sudden death, or at least rapid death. Its frequency is very difficult to analyze. Even a necropsy would sometimes have difficulty differentiating Takotsubo cardiomyopathy from an acute coronary occlusion or a rarer pathology.

Cardiogenic shock may initially complicate Takotsubo cardiomyopathy, and it is important to emphasize that in this case, one must "fight" since we are dealing with a pathology that will completely regress, resorting, if necessary, in extreme cases to transient circulatory assistance.

Many non-specific complications have been described: heart failure, myocardial rupture, aneurysm, rhythm disorders (VT, VF), BAV 3, systemic embolism, mitral insufficiency, right ventricular dysfunction....

The formation of an apical thrombus has been described (Figure 11), with the risk of migration when the left ventricle recovers contraction.

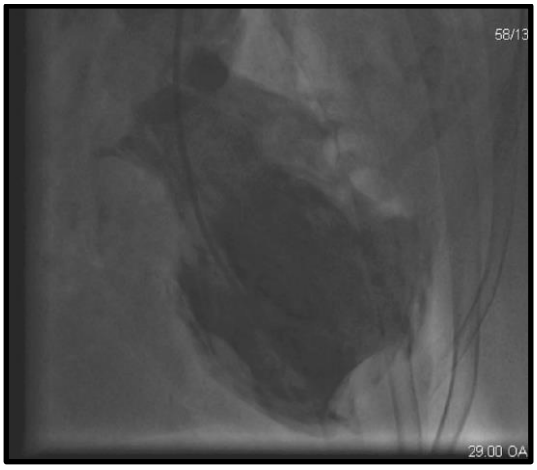

Figure 11: Apical thrombus complicating a severe form of apical Tako-Tsubo

A left intraventricular gradient is present in about 25% of cases, sometimes triggered or aggravated by the use of positive inotropes.

The prognosis of hospitalized forms remains good, however, with a mortality rate of about 3%. A more recent analysis concludes that the rate of complications is higher, probably due to the increasing age of the patients currently diagnosed.

In the vast majority of cases, recovery is complete before the end of the first month. It may be delayed in about 5% of cases. Recurrence is quite rare, about 3.5% at 5 years [18, 19].

Several recent follow-up studies have shown an excess of extracardiac mortality, particularly neoplastic, in the years following takotsubu cardiomyopathy [20].

In the acute phase, the management of patients with Takotsubo cardiomyopathy should be that of an acute coronary syndrome and therefore should be treated as an acute coronary syndrome until coronary lesions are ruled out. Once the diagnosis of Takotsubo cardiomyopathy is confirmed, there is currently no formal recommendation. However, because of this left ventricular dysfunction, prescription of beta-blockers and converting enzyme inhibitors is widely done, at least until recovery ad integrum. As we have seen above, in case of shock without left intraventricular gradient, the prescription of positive inotropic agents (dobutamine) is possible and therapeutic escalation (counter pulsation balloon and circulatory assistance) remains fortunately exceptional. Long-term prescription of beta-blockers to reduce the risk of recurrence is not currently justified, as no study has demonstrated its usefulness.

It is important to make this diagnosis of Takotsubo cardiomyopathy. Indeed, as the clinical presentation is that of an acute coronary syndrome, if the diagnosis is not made, long-term treatment of ischemic heart disease will be instituted. Thus, the patient will be prescribed dual antiplatelet therapy for one year, with the risks of bleeding associated with this unwarranted treatment.

CONCLUSION

Takotsubo cardiomyopathy is an increasingly recognized clinical entity, characterized by apical left ventricular dysfunction with chest pain, electrocardiographic changes, minimal biomarker changes, and no coronary lesions.

In most cases, an episode of emotional or physical stress precedes the event. Its pathophysiological mechanism remains unknown, but high circulating catecholamines seem to be the triggering factor and the most plausible explanation for this primary acquired cardiomyopathy. It is important to be aware of this diagnosis in patients presenting with ACS because its evolution is most often spontaneously favorable.

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