THE ANATOLIAN JOURNAL OF CARDIOLOGY

Official journal of the TURKISH SOCIETY OF CARDIOLOGY

Adrenergic Shock Presenting with Atypical Takotsubo Syndrome: The Alliance Between Levosimendan and Intraaortic Balloon Pump Against the Pheochromocytoma

CASE REPORT

INTRODUCTION

Takotsubo syndrome (TTS) is generally considered benign, nevertheless, hemodynamic instability during the acute phase might expose the patients to a high risk of mortality.[1] Takotsubo syndrome complicated by cardiogenic shock (CS) may, in some rare cases, be caused by a hidden pheochromocytoma.[2] Currently, a standard treatment of adrenergic shock is not established and precise recommendations are not available.

CASE REPORT

A 61-year-old woman with a history of dyslipidemia and smoking was admitted to the Emergency Department for epigastric pain and vomiting. The hemodynamic parameters were normal. Electrocardiogram revealed ST-segment depression in V3-V6 leads (Figure 1). Troponin and myoglobin were elevated, and the arterial blood gas showed metabolic acidosis. Echocardiography revealed an ejection fraction (EF) of 40%, akinesia of basal and mid-left ventricular (LV) segments, and apical hyperkinesia. In addition, a severe mitral regurgitation (MR) was detected. Global longitudinal strain was abnormally low (–14.3%), with impairment mainly in the basal and mid-LV segments (Figure 2; Video 1). Clinical condition deteriorated with hemodynamic instability and anuria. Dobutamine was promptly administered and titrated at 6 µg/kg/min. Emergency coronary angiography showed no significant coronary artery disease. The left ventriculography confirmed an atypical TTS pattern and a severe MR. Due to the dramatic decrease in blood pressure, an intra-aortic balloon pump (IABP) was placed without complications. The patient's clinical condition and the MR immediately improved and, therefore, the dobutamine was titrated off. Levosimendan was started at the dose of 0.05 µg/kg/min. In the following days, markedly high values of transaminases, lactate dehydrogenase, creatinine, amylases, creatinine phosphokinase, suggestive of multiorgan failure were registered. Incidentally, echocardiography extended to the abdominal upper quadrants revealed a right upper pole renal mass. The hypothesis of pheochromocytoma was considered and, in fact, the 24-hour urine catecholamines were measured and found to be strongly elevated. Contrast-enhanced abdominal computed tomography scan showed a solid hypervascular lesion, localized in the right adrenal gland, characterized by intraparenchymal hemorrhagic areas. Intra-aortic balloon pump was removed when hemodynamic parameters became stable. After 10 days, a right adrenalectomy was performed. The histology confirmed the pheochromocytoma (Figure 3). Thereafter, we observed an improvement in the patient's clinical condition, who was discharged completely asymptomatic and showing a full recovery of LV wall motion, EF 60%, and a mild MR.

Maria Vincenza Polito[1]
Elena De Angelis[2]
Andreas Hagendorff[3]
Alessandro Puzziello[4]
Francesco Vigorito[5]
Amelia Ravera[5]

[1]Department of Cardiology, Ospedale Evangelico Betania, Naples, Italy
[2]Division of Cardiology, Department of Medicine, Surgery and Dentistry, University of Salerno, Salerno, Italy
[3]Department of Cardiology, University Hospital Leipzig, Leipzig, Germany
[4]Department of General Surgery, Salerno University Hospital, Salerno, Italy
[5]Department of Cardiology, A.O.U. "San Giovanni di Dio e Ruggi D'Aragona", Salerno, Italy

Corresponding author:
Maria Vincenza Polito
✉ mvpolito@hotmail.it

Cite this article as: Vincenza Polito M, De Angelis E, Hagendorff A, Puzziello A, Vigorito F, Ravera A. Adrenergic shock presenting with atypical takotsubo syndrome: The alliance between levosimendan and intraaortic balloon pump against the pheochromocytoma. Anatol J Cardiol 2022;26(4):325-327.

DOI:10.5152/AnatolJCardiol.2021.522

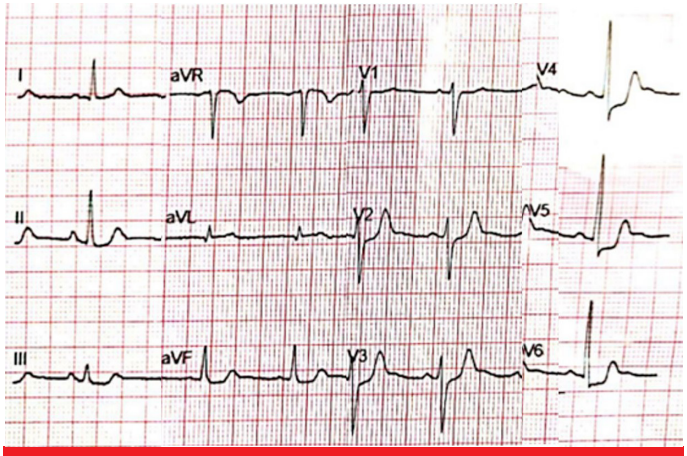

Figure 1. Electrocardiogram at admission showing ST-segment depression in V3-V6 leads.

DISCUSSION

Pheochromocytoma is a recognized cause of secondary TTS.[3] Cardiogenic shock has been described as an initial manifestation in only 2% of pheochromocytoma; this percentage is higher in the presence of intraparenchymal adrenal hemorrhage and/or necrosis because of the transient catecholamine release in the bloodstream, as described in our case.[4]

In the work of Y-Hassan,[5] the patients with pheochromocytoma and TTS were 19 years younger than those without tumors and no precipitating factors were often recognized. Moreover, the complication rate (67.9% vs. 21.8%) and CS (34.2% vs. 4.2%) were higher in patients with pheochromocytoma-induced TTS.[6] The apical ballooning pattern is certainly the most common form of TTS; however, in patients with TTS and pheochromocytoma, an atypical pattern is more frequently identified (30% vs. 2.2% of all comers with TTS).[7]

An important point to consider is that pheochromocytoma-induced CS is difficult to handle as it has been demonstrated that the inotropes may be less effective and be responsible for the worsening of LV wall motion.[8]

Despite the lack of data on the acute phase best strategy in the different TTS patterns, some reports have shown that the Ca^{2+}-sensitizer levosimendan could be safely and effectively used in complicated TTS as an alternative to catecholaminergic agents,[9] is associated with a significant improvement in hemodynamic parameters, a shorter length of hospital stay, and a faster recovery time from TTS.[10]

Unfortunately, there are no previous randomized studies with regard to the use of levosimendan or mechanical circulatory support devices in severe LV dysfunction by shock adrenergic.[11,12]

CONCLUSION

To conclude, in case of atypical TTS complicated by CS, a pheochromocytoma should always be suspected. A prompt diagnosis and hemodynamic stabilization are essential for a good prognosis. Intra-aortic balloon pump support together with levosimendan is valuable and potentially life-saving therapy as a bridge to recovery.

Informed Consent: Written informed consent was obtained from the patient.

Video 1: The top transthoracic echocardiography shows apical 4- and 2-chamber views of the left ventricle (LV). Apical hyperkinesia

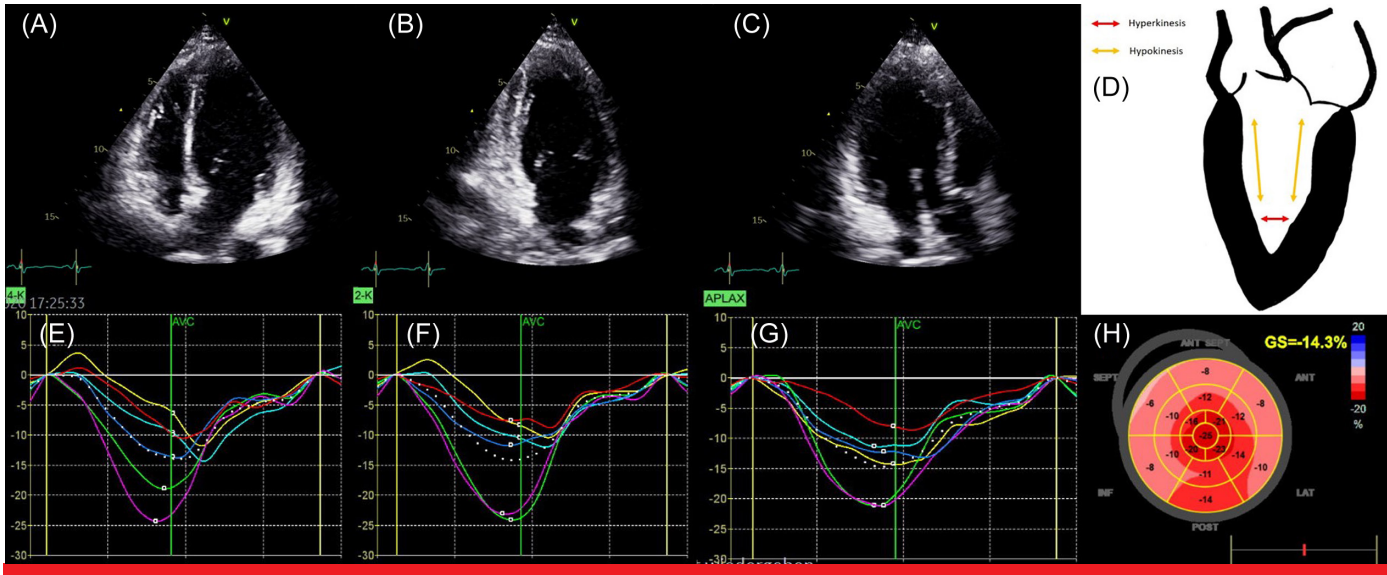

Figure 2. The top transthoracic bidimensional echocardiography shows apical 4- (Panel A), 2- (Panel B), and 3-chamber (Panel C) views of the left ventricle (LV). In this case, apical hyperkinesia associated with hypokinesia of the basal and mid-LV segments (see Panel D at the right) was reported. Below, 2-dimensional speckle-tracking echocardiography analyses for the apical 4- (Panel E), 2- (Panel F), and 3-chamber (Panel G) views. In Panel H, the longitudinal strain bull's eye plot shows a significant reduction of global longitudinal strain (GS) (–14.3%) with impairment mainly in the basal and mid-LV segments.

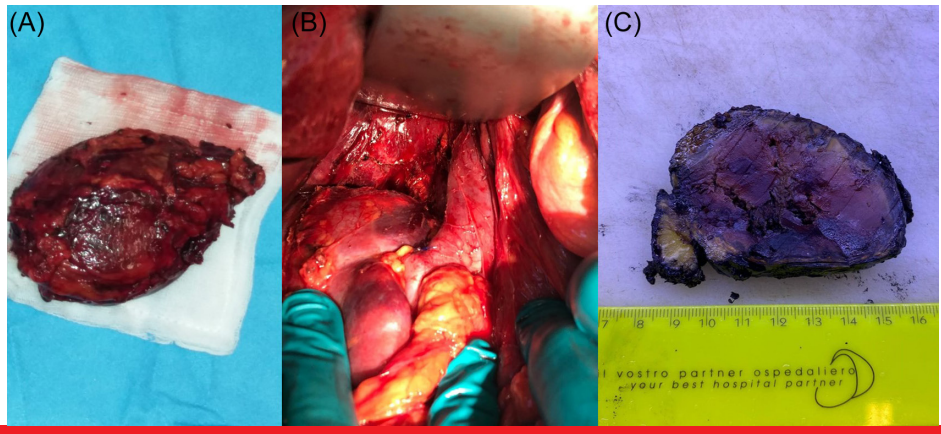

Figure 3. Soft-tissue surrenalic mass with multiple hemorrhagic and necrotic areas (Panel A) was removed from the right surrenal lodge (Panel B) after midline laparotomy incision and opening the layers. The mass was solid, its total size was 7.5 × 6 × 4 cm and weighted 70 g (Panel C). The final histopathological examination confirmed the diagnosis of pheochromocytoma.

associated with hypokinesia of the basal and mid-LV segments is shown. Below, the longitudinal strain bull's eye plot with a significant reduction of global longitudinal strain (GS) and impairment mainly in the basal and mid-LV segments.

REFERENCES

1. Pelliccia F, Pasceri V, Patti G, et al. Long-term prognosis and outcome predictors in takotsubo syndrome: a systematic review and meta-regression study. *JACC Heart Fail*. 2019;7(2):143-154. [CrossRef]
2. Lyon AR, Bossone E, Schneider B, et al. Current state of knowledge on takotsubo syndrome: a position statement from the taskforce on Takotsubo Syndrome of the Heart Failure Association of the European Society of Cardiology. *Eur J Heart Fail*. 2016;18(1):8-27. [CrossRef]
3. Ghadri JR, Wittstein IS, Prasad A, et al. International expert consensus document on takotsubo syndrome (Part I): clinical characteristics, diagnostic criteria, and pathophysiology. *Eur Heart J*. 2018;39(22):2032-2046. [CrossRef]
4. Amin A, Biswas S, Baccay F. Traumatic injury causing intraperitoneal hemorrhage of an occult pheochromocytoma. *Case Rep Crit Care*. 2012;2012:342819. [CrossRef]
5. Y-Hassan S. Clinical features and outcome of pheochromocytoma-induced takotsubo syndrome: analysis of 80 published cases. *Am J Cardiol*. 2016;117(11):1836-1844. [CrossRef]
6. Agarwal V. Takotsubo cardiomyopathy With pheochromocytoma: when an imitator meets a masquerader. *JACC Case Rep*. 2019;1(2):91-93. [CrossRef]
7. Izumo M, Nalawadi S, Shiota M, et al. Mechanisms of acute mitral regurgitation in patients with takotsubo cardiomyopathy an echocardiographic study. *Circ Cardiovasc Imaging*. 2011;4(4):392-398. [CrossRef]
8. Templin C, Ghadri JR, Diekmann J, et al. Clinical features and outcomes of takotsubo (stress) cardiomyopathy. *N Engl J Med*. 2015;373(10) :929-938. [CrossRef]
9. Yaman M, Arslan U, Kaya A, et al. Levosimendan accelerates recovery in patients with takotsubo cardiomyopathy. *Cardiol J*. 2016;23(6):610-615. [CrossRef]
10. Santoro F, Ieva R, Ferraretti A, et al. Safety and feasibility of levosimendan administration in takotsubo cardiomyopathy: a case series. *Cardiovasc Ther*. 2013;31(6) :e133-e137. [CrossRef]
11. Battimelli A, Polito MV, Di Maio M, et al. Stress-related cardiomyopathy, ventricular dysfunction, artery thrombosis: a hidden pheochromocytoma. *Am J Emerg Med*. 2014;32(3):286.e5-286.e9. [CrossRef]
12. Polito MV, Ravera A, Silverio A, et al. A peculiar etiology of acute heart failure: adrenergic myocarditis. *Am J Emerg Med*. 2015;33(10):1545.e3-1545.e6. [CrossRef]