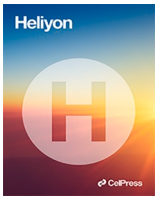
Heliyon 10 (2024) e35337

Contents lists available at ScienceDirect

Heliyon

journal homepage: www.cell.com/heliyon



Case report

A case report of severe left ventricular outflow tract obstruction in a middle-aged adult with chest pain and shock

Chengzhi Chen *, Xiaoyan Yang, Yao Hu

Department of Cardiology, Liuyang People Hospital, Liuyang, 410300, China

ARTICLE INFO

Keywords:
Case report
Left ventricular outflow tract obstruction
Hypertrophic obstructive cardiomyopathy
Left ventricular apical ballooning
Endocardial radiofrequency ablation of septal hypertrophy

ABSTRACT

Severe left ventricular outflow tract obstruction (LVOTO) of hypertrophic cardiomyopathy is an acutely life-threatening, must-not miss, cardiology emergency that infrequently presents to the emergency department (ED). Patients with this condition usually manifest chest pain, syncope, cardiogenic shock, and severe ischemia. LVOTO is easy misdiagnosed as acute coronary syndrome. In our patient, the ECG showed a significant ST-segment depression and a 0/0 mmHg blood pressure when the peak left ventricular outflow tract gradient was abruptly increased by provocable activities. However, the patient had normal coronaries on cardiac catheterization, and, upon being immediately treated with intravenous esmolol, his symptoms were relieved and blood pressure was normal after 30 minutes. This case highlights, not only that early and exact diagnosis of LVOTO is crucial, but also the importance of the therapeutic strategies used.

1. Background

Severe left ventricular outflow tract obstruction (LVOTO) is the major cause of severe cardiogenic shock of hypertrophic obstructive cardiomyopathy (HOCM). LVOTO patients usually manifest with features such as an acute coronary syndrome and cardiogenic shock, which is different from shock resulting of other causes, also requiring a different treatment. Some of these patients have shown ST segment change, but, like in our case, diffuse ST-segment depression and shock were relatively rare [1].

2. Case presentation

A 52-year-old man was admitted to the emergency department with chest pain, profuse sweating, and dyspnea, due to intense arguing with others. He had a 5-year history of recurrent chest pain and syncope and no relevant medical history. His blood pressure was 0/0 mmHg and showing no systolic ejection murmurs of the sternum left border. The ECG was obtained (Fig. A), showing a prominent ST depression (STD) in leads V_5–V_6, II, and aVF, and ST elevation (STE) occurred in leads aVR and sinus bradycardia. Transthoracic echocardiography (TTE) showed HOCM with mitral valve systolic anterior motion (SAM) (Fig. B) and a left ventricular outflow tract (LVOT) peak pressure gradient of 172.1 mmHg. TTE demonstrated no regional wall motion abnormality and the LV ejection fraction was normal (Video A), but there is a decrease in diastolic function. ECG changes were consistent with a severe left ventricular outflow tract obstruction. After an immediate intravenous treatment with esmolol (a 100 mg bolus followed by continuous infusion at 200 μg/kg/min), his symptoms were relieved and the blood pressure was normal after 30 minutes. A subsequent TTE

* Corresponding author.
E-mail address: chengzhiou@163.com (C. Chen).

https://doi.org/10.1016/j.heliyon.2024.e35337
Received 4 April 2024; Received in revised form 7 July 2024; Accepted 26 July 2024
Available online 3 August 2024

showed that the pressure gradient of the LVOT was 31.4 mmHg. Follow-up ECG showed a baseline regression of the ST segment displacement. Cardiac troponin T (cTnT) and N-terminal pro−b-type natriuretic peptide levels were 24.45 pg/mL (reference: <14 pg/mL) and 455.9 pg/mL (reference: <125 pg/mL), respectively. The patient received 47.5 mg oral metoprolol succinate and 90 mg verapamil daily. Coronary angiography revealed normal coronary arteries. A simultaneous pressure gradient in the LVOT was measured and the LVOT gradient was 60.2 mmHg at rest. Cardiac magnetic imaging (CMR) was suggestive of hypertrophic obstructive cardiomyopathy and showed areas of LGE in the septal, inferior LV segment. The patient rejected septal myectomy or alcohol ablation and received endocardial radiofrequency ablation of septal hypertrophy (ERASH). The patient received ERASH guided by intracardiac echocardiography (ICE) (Video B). After the procedure, the LVOT gradient was 10 mmHg and SAM of the mitral valve was still detetable (Video C). More than 4 hours after radiofrequency, the patient experienced again chest pain, profuse sweating, shortness of breath, and a blood pressure of 0/0 mmHg. Repetition of the ECG showed a similar result to the first ECG, but with a wider QRS wave than the first ECG. Immediate intravenous injection of 10 mg dexamethasone and 100 mg esmolol did not alleviate the above-mentioned symptoms. Shortly after, the patient had a cardiac arrest, cardiopulmonary resuscitation failed, and the patient died.

3. Discussion

Hypertrophic cardiomyopathy (HCM) is defined as an LV wall thickness ≥15 mm in any myocardial segment (not solely explained by loading conditions). When the peak LVOT gradient is ≥ 30 mm Hg, HOCM is diagnosed. HCM and severe aortic stenosis are the two most common diseases in patients with chest pain, syncope, and systolic murmurs. However, the diagnosis was confirmed by TTE. The final diagnosis was HOCM. The degree of LVOTO of HOCM is dynamic and dependent on ventricular load and contractility. The association between obstruction severity and cardiovascular symptoms (including chest pain and sudden death) has been identified. The dynamic nature of the obstruction challenges independent risk predictor detection [1–3].

When the LVOT gradient was abruptly increased by provocable activities, severe LVOTO suddenly developed. These patients manifested chest pain, severe ischemia cardiogenic shock and syncope· ECG abnormalities were the following: ST-segment elevation, ST depression, or diffuse T-wave inversion [4–6]. Mechanistically, acute severe LVOTO results in the obstruction of forward blood flow and in hemodynamic collapse, which leads to a severe decrease in coronary perfusion and oxygen supply. Furthermore, LVOTO is associated with a high left ventricular systolic pressure and increased oxygen consumption. A mismatch between oxygen supply and its demand can result in extensive subendocardial ischemia, leading to diffuse and obvious ST-segment changes [1,3,4].

When chest pain and ischemic ECG abnormalities are the main HOCM symptoms, they can be easily misdiagnosed as acute coronary syndrome and treated with coronary vasodilators, such as nitroglycerin. When dyspnea is the main clinical manifestation, it can be easily misdiagnosed as acute left heart failure and treated with positive inotropic agents and vasodilation. However, for HOCM patients, positive inotropic agents and vasodilation are contraindicated and may even have potentially catastrophic effects. Therefore, early diagnosis is crucial.

Importantly, shock caused by severe LVOTO treatment differs from shock resulting from other causes. The therapeutic mainstay is to relieve obstruction, being intravenously administered β-blocker therapy the most important therapeutic step. It may seem counterintuitive to treat patients with β-blockers when their BP is low; however, in our experience, this strategy shows that LVOT gradients often decrease, and BP increases. When, despite optimal medical therapy, the patients remain significantly symptomatic, mechanical circulatory support (like VA-ECMO) and the surgical relief of LVOTO are required [1,4,7].

As reported, 0.9 % HOCM patients manifested an acute episode of left ventricular apical ballooning (LVAB) when they had high LVOT gradients [1]. LVAB is caused by the sudden onset of a latent LVOTO [1,4,7]. However, there were also some HOCM patients without any acute episodes of LVAB when they had LVOTO [5]. Furthermore, apical-variant hypertrophic cardiomyopathy patients without LVOTO also had LVAB [8]. The essence of LVAB is considered to be a paradoxical reversible systemic dysfunction [4]. The precise mechanism of LVAB in patients with HOCM and acute LVOTO is unclear. In our case, we did not observe any apical balloon dilation, which may be related to a timely relief of LVOTO.

In the present study, the association between the severity of the obstruction and the predisposition to sudden death was demonstrated. LVOTO requires an aggressive therapy to eliminate the obstruction, which initially includes pharmacological intervention, mechanical circulatory support, alcohol septal ablation, and septal myectomy. Compared with ablation, which may require weeks for effect, surgery provides immediate and reliable relief of the obstruction. For these reasons, surgery is recommend relative to alcohol ablation [4,7]. Our patient received ERASH guided by ICE, which is a new treatment option for HOCM [9]. Postoperative chest pain and the repeated ECG indicated that the patient had acute dynamic LVOTO again, despite not being verified by TTE. The mechanism was still not clear, and an acute edema after radiofrequency ablation could be a potential cause [10]. It is established that ERASH cannot immediately and reliably relief obstruction. According to our experience, postoperative death of an ERASH patient like ours has been rarely published worldwide. The outcome suffered by our patient indicates that the safety and indication of ERASH still need further research, especially for patients with acute LVOTO.

Article information

Department of Cardiology, Liuyang People Hospital, No. 452 Daowushan west Rd,Liuyang, 410300, (chengzhiou@163.com) Tel.: +86-0731-83647063 Fax: +86-0731-68313019.

Ethics statement

All authors have signed forms declaring no conflict of interest. The informed consent was obtained from family members of the deceased patient for publication.

Data availability statement

Data will be made available on request.

Funding/support

The work was supported in part by grants from Medical Scientific Research Foundation of Hunan Province (no. B202303016199) and the Research Foundation of Science and Technology Bureau of Liuyang City, China (no. K20011040).

Role of the funder/sponsor

The funders had no role in the preparation, review, or approval of the manuscript and decision to submit the manuscript for publication.

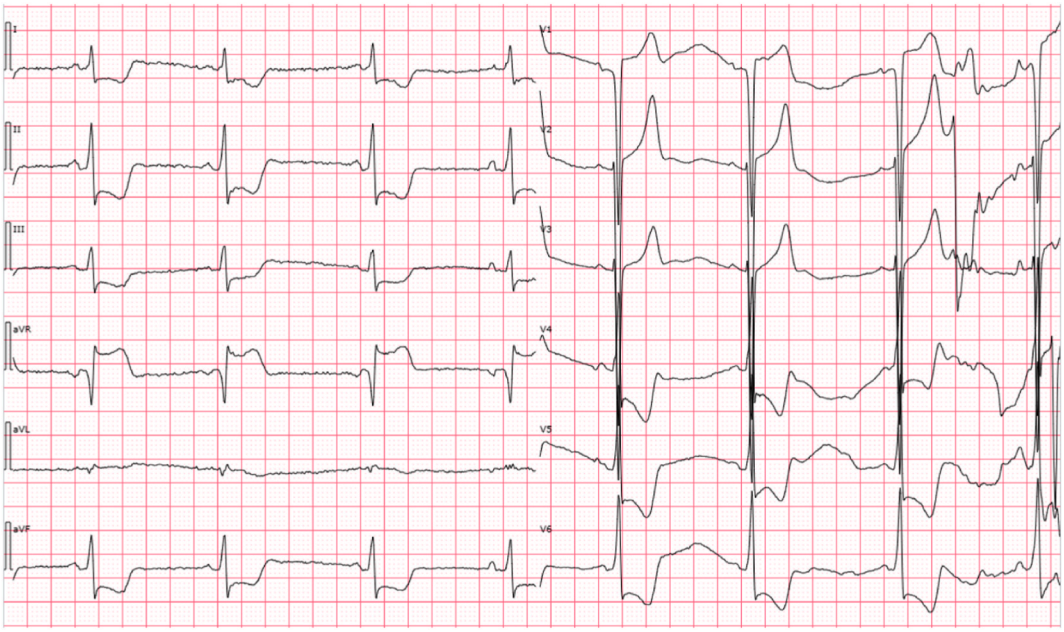


Fig. A. ECG obtained after he argumented with others and felt chest pain, his Blood pressure was 0/0 mmHg. A TTE showed mitral valve systolic anterior motion (SAM) and peak pressure gradient in LVOT of 172.1 mmHg. The ECG showed bradycardia, marked ST-segment elevation in the aVR and diffuse ST-segment depression.

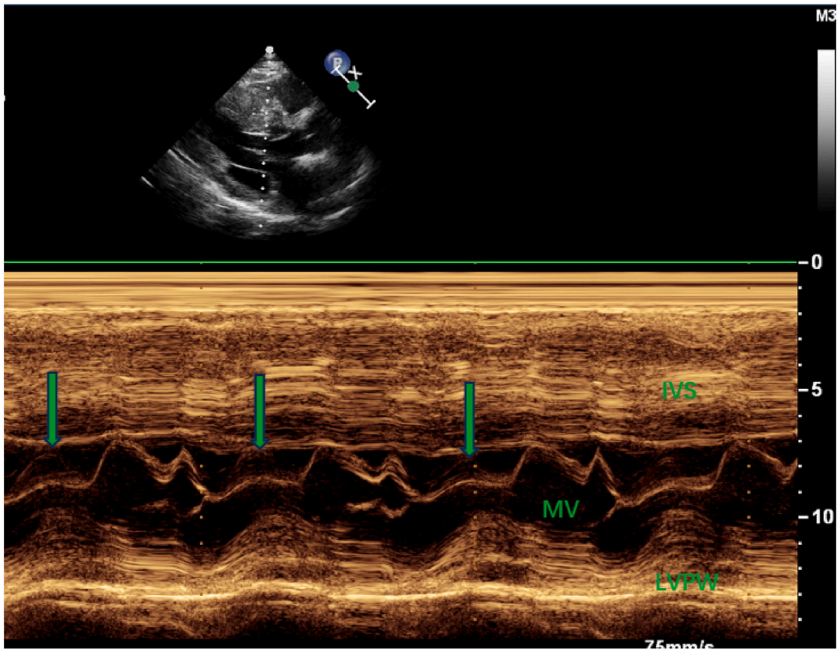


Fig. B. M-mode echocardiograms recorded in our patient with hypertrophic cardiomyopathy demonstrating disproportionate septal hypertrophy and SAM of the mitral valve (arrow).

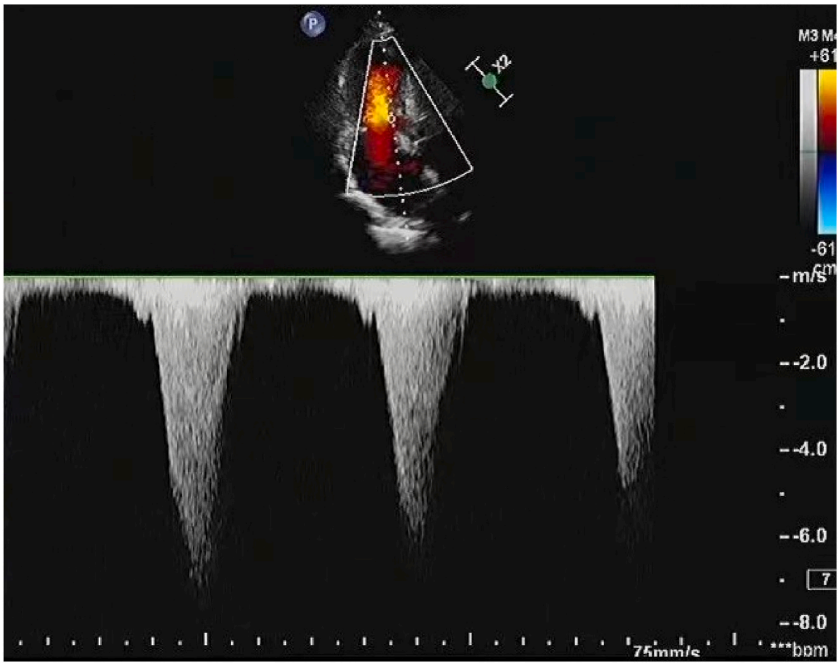


Fig. C. A TTE showed mitral valve systolic anterior motion (SAM) and peak pressure gradient in LVOT of 172.1 mmHg.

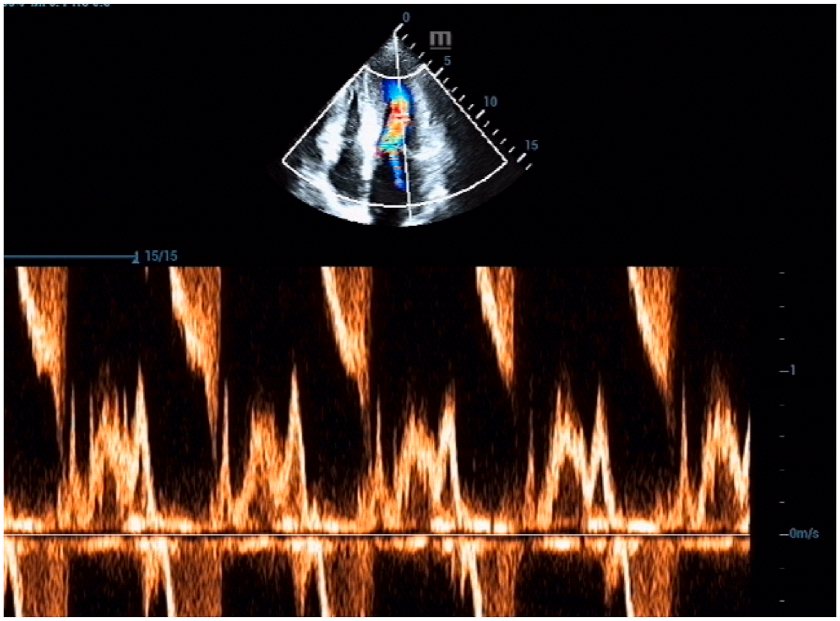


Fig. D. A TTE showed a decrease in diastolic function.

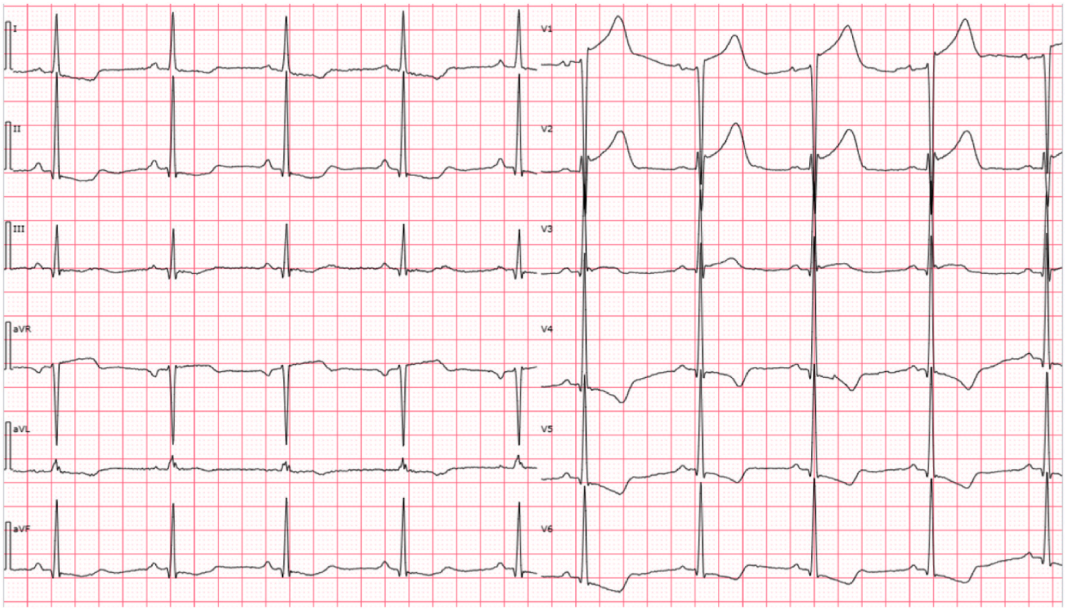


Fig. E. ECG obtained 32 minutes after the first ECG, his blood pressure was recovered. Subsequent TTE indicated a 31.4 mmHg LVOT pressure gradient.The ECG showed basely regression of the ST segment displacement.

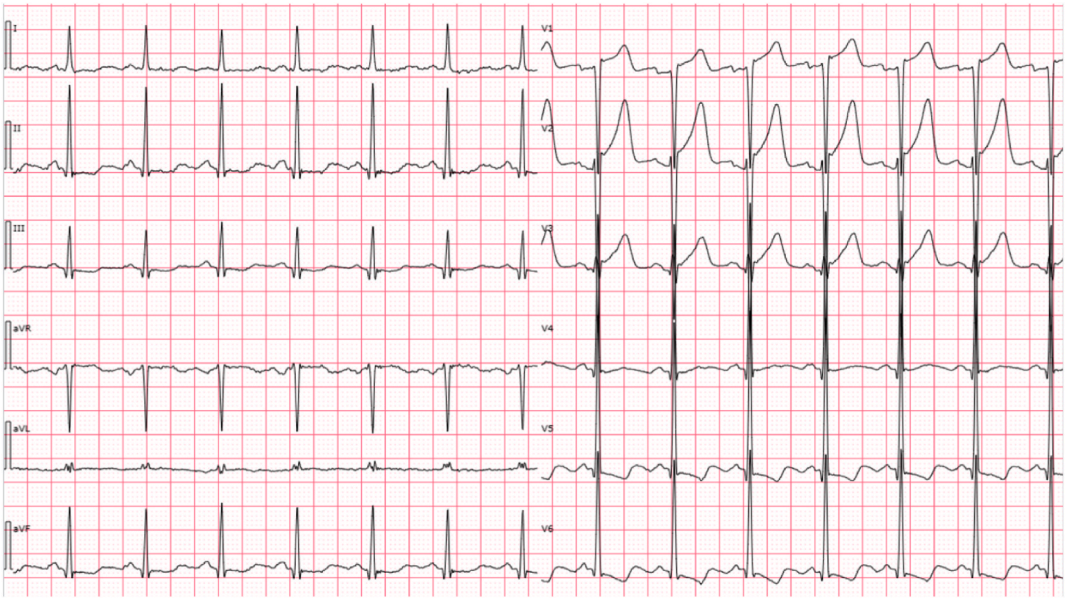


Fig. F. ECG on the second day after admission, the patient without any discomfort symptoms.Transthoracic echocardiography (TTE) showed a LVOT peak pressure gradient of 57.8 mmHg. The ECG showed a sinus rhythm at a rate of 94 beats per minute with ST elevation (STE) in leadsV1 toV3 (pseudo-STEMI pattern), ST depression (STD) in leads V5 to V6, II, and aVF, deep narrow Q waves in the lateral (V5,V6) and inferior (II, III, and aVF) leads, Notable LVH by voltage criteria(modified Sokolow-Lyon score ≥50 mm), P wave duration is 140 ms, corrected QT interval (Bazett's formula) 520 ms, fragmented QRS in leads V3, II, III, and aVF.

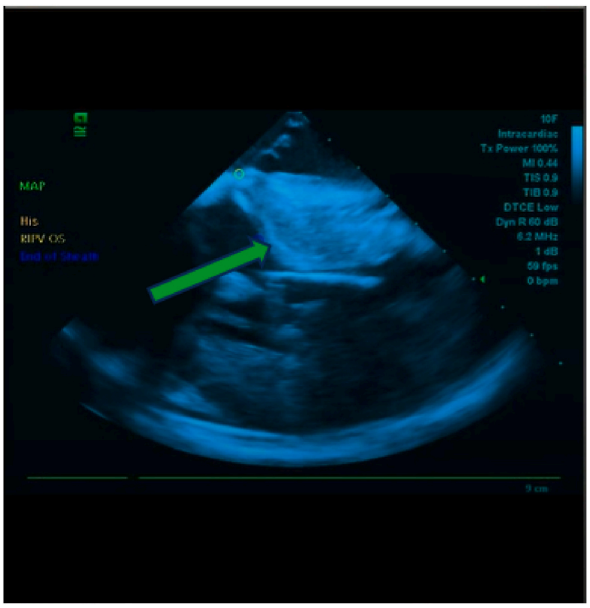


Fig. G. ICE showed acute edema after radiofrequency ablation (arrow).

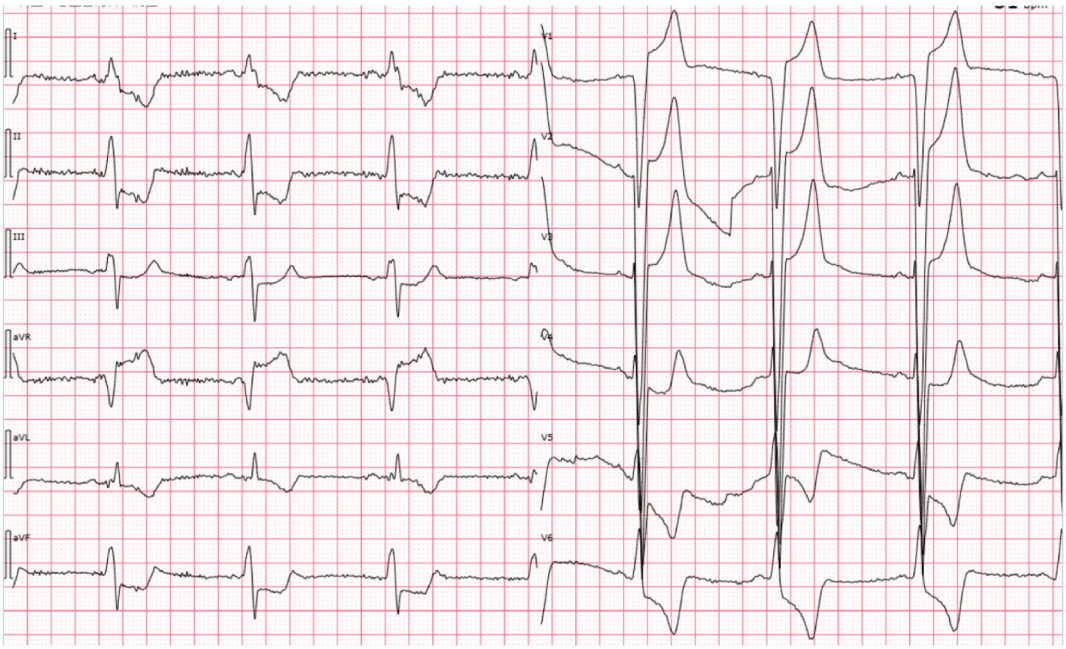


Fig. H. ECG obtained 4 hours postoperative when the second shock occurred, shortly after, the patient died of sudden cardiac arrest. The ECG showed bradycardia, QRS duration 150 ms, complete left bundle branch block, marked ST-segment elevation in the aVR and diffuse ST-segment depression.

CRediT authorship contribution statement

Chengzhi Chen: Writing – original draft, Writing – review & editing. **Xiaoyan Yang:** Writing – review & editing. **Yao Hu:** Writing – review & editing.

Declaration of competing interest

The authors have no conflicts of interest to declare.

Appendix A. Supplementary data

Supplementary data to this article can be found online at https://doi.org/10.1016/j.heliyon.2024.e35337.

References

[1] M.V. Sherrid, K. Riedy, B. Rosenzweig, et al., Hypertrophic cardiomyopathy with dynamic obstruction and high left ventricular outflow gradients associated with paradoxical apical ballooning, Echocardiography 36 (1) (2019 Jan) 47–60, https://doi.org/10.1111/echo.14212.
[2] e Mattos B. Piva, M.A. Torres, T.F. Rebelatto, et al., The diagnosis of left ventricular outflow tract obstruction in hypertrophic cardiomyopathy, Arq. Bras. Cardiol. 99 (1) (2012 Jul) 665–675, https://doi.org/10.1590/s0066-782x2012000700013. English, Portuguese.
[3] R.A. Nishimura, H. Seggewiss, H.V. Schaff, Hypertrophic obstructive cardiomyopathy: surgical myectomy and septal ablation, Circ. Res. 121 (7) (2017 Sep 15) 771–783, https://doi.org/10.1161/CIRCRESAHA.116.309348.
[4] M.V. Sherrid, D.G. Swistel, I. Olivotto, et al., Syndrome of reversible cardiogenic shock and left ventricular ballooning in obstructive hypertrophic cardiomyopathy, J. Am. Heart Assoc. 10 (20) (2021 Oct 19) e021141, https://doi.org/10.1161/JAHA.121.021141.
[5] Y. Harano, Y. Kawase, H. Matsuo, Dynamic improvement of an acute exacerbated subaortic pressure gradient after intravenous propranolol and cibenzoline, recorded using a pressure wire: a case report, Eur Heart J Case Rep 6 (8) (2022 Jul 25) ytac311, https://doi.org/10.1093/ehjcr/ytac311.
[6] E. Biagini, C. Pazzi, I. Olivotto, et al., Usefulness of electrocardiographic patterns at presentation to predict long-term risk of cardiac death in patients with hypertrophic cardiomyopathy, Am. J. Cardiol. 118 (3) (2016 Aug 1) 432–439, https://doi.org/10.1016/j.amjcard.2016.05.023.
[7] R. Citro, M. Bellino, E. Merli, et al., Obstructive hypertrophic cardiomyopathy and Takotsubo syndrome: how to deal with left ventricular ballooning? J. Am. Heart Assoc. 12 (21) (2023 Nov 7) e032028 https://doi.org/10.1161/JAHA.123.032028.
[8] R.R. Roy, F.A. Hakim, R.T. Hurst, et al., Two cases of apical ballooning syndrome masking apical hypertrophic cardiomyopathy, Tex. Heart Inst. J. 41 (2) (2014 Apr 1) 179–183, https://doi.org/10.14503/THIJ-13-3191.
[9] T. Lawrenz, B. Borchert, C. Leuner, et al., Endocardial radiofrequency ablation for hypertrophic obstructive cardiomyopathy: acute results and 6 months' follow-up in 19 patients, J. Am. Coll. Cardiol. 57 (5) (2011 Feb 1) 572–576, https://doi.org/10.1016/j.jacc.2010.07.055.
[10] T. Lawrenz, D. Lawin, K. Radke, et al., Acute and chronic effects of endocardial radiofrequency ablation of septal hypertrophy in HOCM, J. Cardiovasc. Electrophysiol. 32 (10) (2021 Oct) 2617–2624, https://doi.org/10.1111/jce.15203.