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**CASE REPORT**
*Cardiovascular imaging*

# A case report of left ventricular outflow tract obstruction due to early growth of a calcified amorphous tumour despite normal renal function

**Misayo Hayashi [1]*, Takayoshi Kato[2], Keita Fujimoto[3], Takatomo Watanabe[1], and Hiroyuki Okura[1]**

[1]Department of Cardiology, Gifu University Graduate School of Medicine, 1-1 Yanagido, Gifu 501-1194, Japan; [2]Department of Cardiovascular Surgery, Gifu University Graduate School of Medicine, 1-1 Yanagido, Gifu 501-1194, Japan; and [3]Department of Radiology, Gifu University Graduate School of Medicine, 1-1 Yanagido, Gifu 501-1194, Japan

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**Background** Calcified amorphous tumours (CAT) are non-neoplastic cardiac masses that have been the focus of several recent studies. Moreover, CAT is frequently observed in women and in patients undergoing dialysis.

**Case summary** A woman in her 70 s with normal renal function was referred to our hospital with a chief complaint of shortness of breath upon effort. Echocardiography and contrast-enhanced CT revealed a cardiac mass with calcification in the intervalvular fibrosa (IVF) of the anterior mitral valve, resulting in left ventricular outflow tract (LVOT) obstruction. One year later, the cardiac mass expanded, and the LVOT obstruction worsened. The patient underwent surgical resection of the mass and double-valve replacement with reconstruction of the IVF (commando operation) and myectomy. Histological examination confirmed that the mass was CAT.

**Discussion** We encountered a patient who underwent valve replacement and a commando operation due to rapidly progressive CAT and consequent progression of LVOT obstruction despite normal renal function. In patients with normal renal function, factors other than calcium and phosphate metabolism contribute to the formation of CAT. Understanding the accumulation of CAT is crucial for understanding its pathogenesis.

**Keywords** Case report • Calcified amorphous tumour (CAT) • Left ventricular outflow tract (LVOT) • Mitral annular calcification (MAC) • Commando operation

**ESC curriculum** 6.8 Cardiac tumours • 2.2 Echocardiography • 2.4 Cardiac computed tomography • 2.1 Imaging modalities • 7.5 Cardiac surgery

## Learning points

- CAT is frequently observed in women and patients undergoing dialysis, but this patient had normal renal function. Normally, if CAT is found, it does not expand or expands slowly over time. However, in this case, the CAT expanded within a year, and LVOT obstruction worsened.
- Resection is the preferred surgical method, with valve replacement occasionally considered.
  In this case, we chose surgical resection of the tumour and double-valve replacement with a reconstruction of the IVF (Commando operation) and myectomy. The Commando operation has recently been accepted for complex infective endocarditis, particularly in cases involving both the aortic and mitral valves and the IVF structure.

* Corresponding author. Tel: +81-058-230-6523, Email: misa.h.0214@gmail.com
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## Background

A calcified amorphous tumour (CAT) is a non-neoplastic mass originally proposed in 1997.[1] Pathologically that is a chronic inflammation of degenerated serum components accompanied by calcification. Although several case reports have been published, the detailed pathogenesis of CAT remains unclear. In some cases, mitral annular calcification (MAC)-related CAT has been documented mainly in patients with end-stage renal disease. Here, we present a case of MAC-related CAT in a patient with normal renal function.

## Summary figure

| Time | Event |
|---|---|
| Initial presentation | A woman in her 70 s with normal renal function visited a clinic for rehabilitation due to lumbar spinal canal stenosis. She reported shortness of breath on exertion. Transthoracic echocardiography (TTE) performed at the clinic revealed a calcified mass in the intervalvular fibrosa (IVF) of the anterior mitral leaflet (AML). |
| Day 0 | The patient was referred to our hospital for further examination. A calcified tumour was also detected by TTE in the IVF of the AML, and we found the calcified mass and hypertrophy of the ventricular septum lead left ventricular outflow tract (LVOT) obstruction. |
| 4 months | We performed first follow-up TTE. The LVOT obstruction and CAT size remained unchanged. We prescribed a β-blocker, which was bisoprolol 1.25 mg. |
| 8 months | We performed second follow-up TTE. TTE showed that the pressure gradient increased with the Valsalva manoeuvre. Since the subjective symptoms remained unchanged, we only increased the dose of the beta blocker. |
| 12 months | Patient experienced dizziness and light-headedness. CT showed that the calcified tumour was expanding in the IVF, and the calcification seemed to extend to NCC. |
| 13 months | Patient was admitted and underwent catheterization. The mean and maximal pressure gradient of the LVOT after the Valsalva manoeuvre were 31 and 51 mmHg, respectively. We tested nitrogen provocation, and the maximum pressure gradient of the LVOT increased to 75 mmHg. The NT-proBNP level increased to 915 pg/mL. |
| 14 months | Surgical resection of the tumour and double-valve replacement with a reconstruction of the IVF (Commando operation) and myectomy underwent. |
| 16 months | It took some time to manage post-operative sick sinus syndrome, but she was discharged without any other complications. Patient was rehabilitated and discharged our hospital. |
| After discharge from hospital | We have continued regular outpatient follow-up of the patient, who has remained stable with rehabilitation, with no recurrence of CAT in TTE. |

## Case presentation

A woman in her 70 s with a medical history of non-tuberculous mycobacterial disease was referred to our hospital with exertional dyspnoea and an abnormal cardiac mass. She was under follow-up observation with expectorant medication more than 2 months when she experienced exertional dyspnoea. Her heart rate was 59 beats per minute, and her blood pressure was 158/101 mmHg, which was elevated at the time of presentation. Subsequent blood pressure measurements were within the normal range. Physical examination revealed no jugular vein distension. Breath sounds were clear, with no rales. Heart sounds were regular, and a systolic murmur graded as Levine II/VI was auscultated at the second intercostal space on the right sternal border. She did not have leg oedema. Transthoracic echocardiography (TTE) detected a calcified tumour of the intervalvular fibrosa (IVF) of the anterior mitral leaflet (AML), leading to left ventricular outflow tract (LVOT) obstruction (LVOTO). (*Figure 1*, Supplementary material online, *Video S1*) A follow-up TTE revealed normal left ventricular function with an ejection fraction of 60%, mitral stenosis (MS) due to a calcified tumour, and MAC (calcified or non-rheumatic MS).

The mitral valve (MV) area was 1.39 $cm^2$, the peak transmitral flow velocity was 1.5 m/s, and the mean pressure gradient (PG) was 2.4 mmHg. Mild MS was observed. Mitral regurgitation was trivial. The tumour size was 12 × 12 mm, and the peak PG of the LVOT was 21 mmHg at rest. The tumour appeared smooth, and she did not show any signs or symptoms of embolization. Transesophageal echocardiography (TEE) displayed calcifications from A2 to A3 of the MV leaflets. Computed tomography (CT) displayed a calcified AML tumour. Her N-terminal pro-brain natriuretic peptide (NT-proBNP) level was 311 pg/mL (normal < 125 pg/mL); C-reactive protein (CRP) levels were 0.07 mg/dL (normal≦0.14 mg/dL), fibrinogen levels were 386 mg/dL(normal range 200–400 mg/dL), serum calcium levels were 9.2 mg/dL (normal range 8.8–10.1 mg/dL), and her renal function was almost normal (creatinine was 0.77 mg/dL (normal range 0.46–0.79 mg/dL) and estimated glomerular filtration rate from creatinine was 55.7 mL/min/1.73 $m^2$ (normal≧60 mL/min/1.73 $m^2$). Electrocardiography revealed a first-degree atrioventricular block. Based on these findings, conservative treatment and regular TTE follow-ups were planned.

One year later, the patient experienced dizziness and light-headedness. The tumour size was 15 mm × 12 mm, and the peak PG

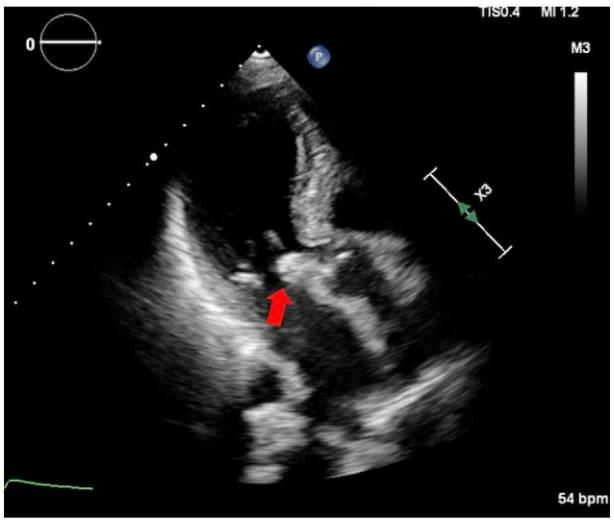


**Figure 1** Transthoracic echocardiogram. A calcified tumour detected at the anterior mitral leaflet and left ventricular outflow tract obstruction in the apical three-chamber view.

of the LVOT was 78 mmHg at rest and 85 mmHg after the Valsalva manoeuvre. CT confirmed that the calcified tumour was expanding during IVF, and the calcification seemed to extend to the non-coronary cusp (NCC) (*Figure 2*, Supplementary material online, *Video S2*). Cardiac catheterization revealed no significant coronary artery stenosis. The mean and maximal LVOT PGs after the Valsalva manoeuvre were 31 and 51 mmHg, respectively (*Figure 3*). Nitrogen provocation was tested and the maximum PG of the LVOT increased to 75 mmHg. The Brockenbrough phenomenon was observed during catheter examination, and the maximum PG of the LVOT increased to 145 mmHg. (*Figure 4*) The peptide level increased to 915 pg/mL. Cibenzoline (300 mg/day) and beta-blockers (bisoprolol, 5 mg/day) were administered. However, no significant reduction was observed in the PG of LVOTO. Therefore, we recommended surgical resection of the tumour, double-valve replacement with IVF reconstruction (the Commando operation), and septal myotomy. Under conventional cardiopulmonary bypass and aortic clamping, we performed a transseptal atriotomy and extended the aortotomy by opening the IVF (*Figure 4A*).[2] Narrowing of the LVOT due to the hypertrophic ventricular septum and a protruding tumour was observed. The tumour originated mainly in the right fibrous trigone and spread to the aortic and mitral annuli. The tumour surface was smooth and covered the endocardium (*Figure 4B*). However, radical tumour resection with preservation of both native valves is impossible because of the impaired area. After myectomy, the aortic cusps and AMLs were removed. To prevent CAT recurrence, it was completely resected using an ultrasonic surgical aspirator, and the fibrous tissue defect was covered with bovine pericardium (Edwards Lifesciences LLC, Irvine, CA, USA). Commando Operation was performed using a 22 mm mechanical valve (Medtronic Inc.; Columbia Heights, MN, USA) for the aortic valve and a 25 mm mechanical valve (St. Jude Medical, St. Paul, MN, USA) for the MV, along with a bovine pericardium (Edwards Lifesciences LLC; Irvine, CA, USA) for reconstructed IVF. We used mechanical prostheses to avoid disturbing LV outflow because the cardiac chamber was relatively small. On gross examination, the tumour was surrounded by fibrous tissues, firm with calcification and debris, and measured 1.4 × 0.8 cm (*Figure 4C*). Histopathological examination revealed calcium and eosinophilic deposits within the cavities. Macrophage infiltration was observed in the background (*Figure 4D*). These findings indicated that the tumour was a CAT.

Post-operative TTE showed that the CAT had been successfully removed, and LVOTO was improved with a significantly better LVOT PG during the Valsalva manoeuvre compared to preoperative levels (*Figure 5*, Supplementary material online, *Video S3*). Managing post-operative sick sinus syndrome took some time, but the patient was discharged without any other complications. There was no recurrence of CAT or other symptoms after surgery. At her most recent follow-up, one and a half years post-discharge, she has been visiting the referring clinic once a month and attending rehabilitation three to four times per month.

## Discussion

The clinical features and current management and prognosis of CAT were described in a systematic review by de Hemptinne *et al.*[3] According to the review, the mean age at presentation was 54 years, with a predominance of women (64%). Although CATs were detected in all the cardiac chambers, but was most prevalent in the MV or annulus (36%). The most frequent symptoms included dyspnoea, syncope, and embolization. The most frequently associated conditions were valve disease (31%), concomitant MAC (14%), end-stage renal disease (ESRD) (21%), diabetes (14%), and coronary artery disease (12%).[3] As ESRD causes metabolic calcium and phosphorus disorders, ectopic calcification occurs in the vascular walls and other areas, contributing to rapid growth and pathological changes.[4,5] Most MAC-related cases with CAT have been reported in patients with ESRD patients; however, the CAT in our case was enlarged despite normal renal function.

Surgical treatment of patients with CAT is often indicated for those at a high risk of embolization. Resection is the preferred surgical method, although valve replacement is occasionally considered. In a review of case reports, 64% of patients with CAT underwent only resection, 16% underwent MV replacement, 10% underwent double-valve replacement and 10% underwent mitral valvuloplasty.[6] In this case, the CAT rapidly expanded in 1 year and CT and TTE revealed calcification extending to the NCC and IVF suggesting progression to aortic valve stenosis. Therefore, it was determined that complete resection of the calcified IVF with valve replacement is required to prevent aortic valve stenosis.

Commando Operation has recently been accepted for complex infective endocarditis, particularly in cases involving both the aortic and MVs and the IVF structure. It is also applied in patients with small annuli requiring double-valve replacement.[2] When infection or calcification spreads from IVF to AML, both valves are resected using the method described by Manouguian *et al.*[7] as a method of valve ring enlargement for two-valve replacement. After the infected area was well dissected, a two-valve replacement was performed, and the intervalve and aortic valve incision lines and left atrial ceiling were reconstructed by patching. We performed tumour resection, double-valve

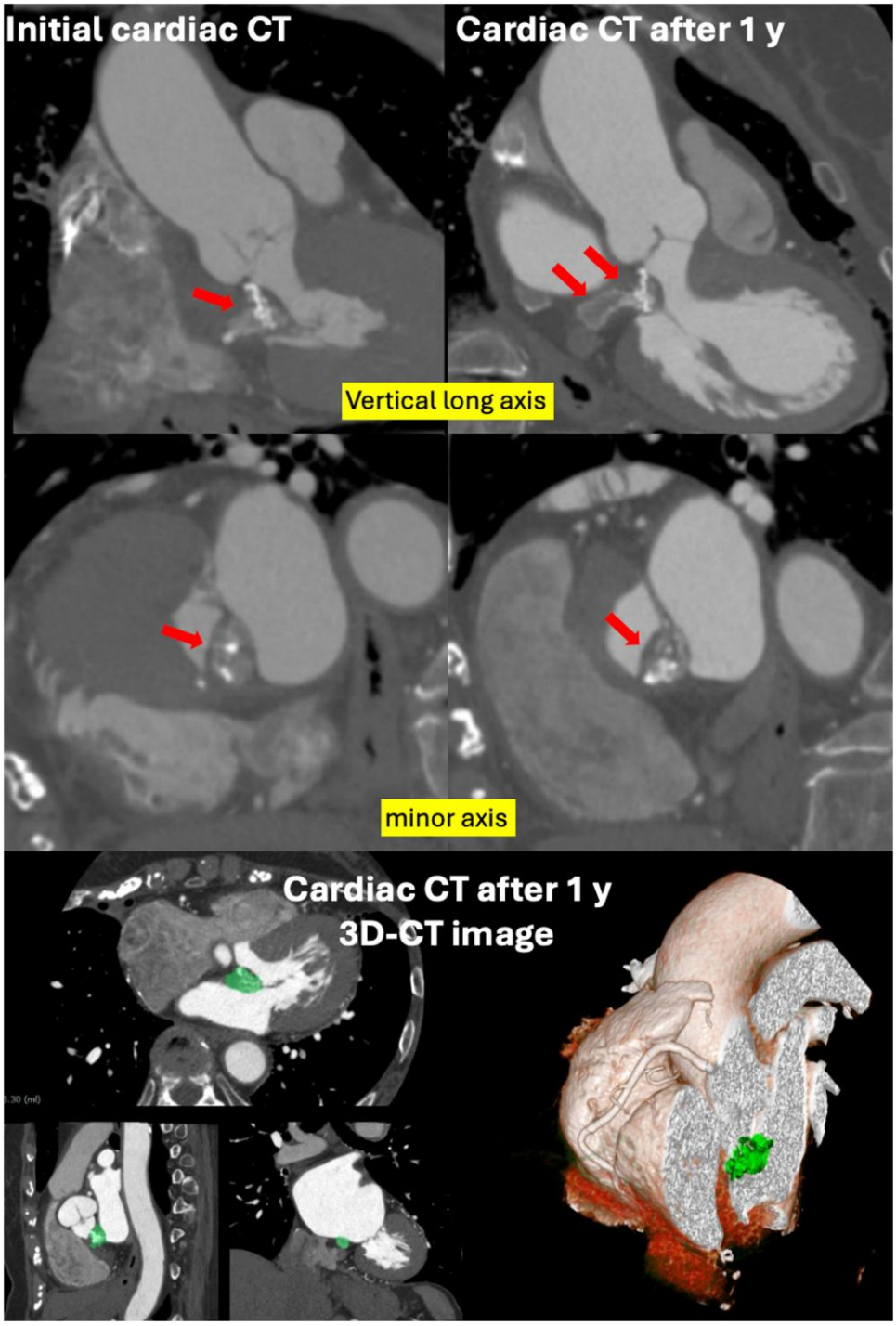


**Figure 2** Cardiac computed tomography displays a calcified mass. The mass seems to extend to the posterior of the left atrium and non-coronary cusp.

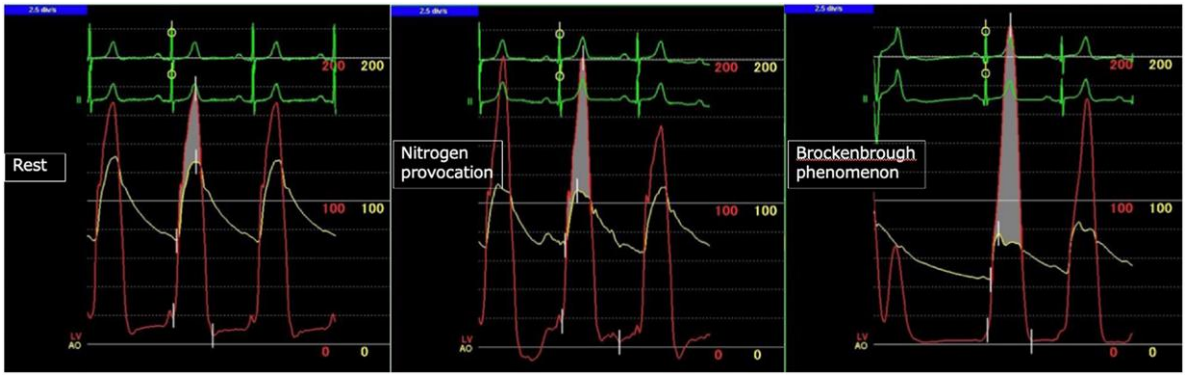


**Figure 3** In pressure measurement with a catheter, the mean pressure gradient of the left ventricular outflow tract with valsalva provocation is 31 mmHg, and the maximum pressure gradient is 51 mmHg. We tested nitrogen provocation using nitro-glycerine (100 μg), displaying 75 mmHg in the maximum pressure gradient of the left ventricular outflow tract. When the Brockenbrough phenomenon was observed during catheter examination, the maximum pressure gradient of the left ventricular outflow tract was rising to 145 mmHg.

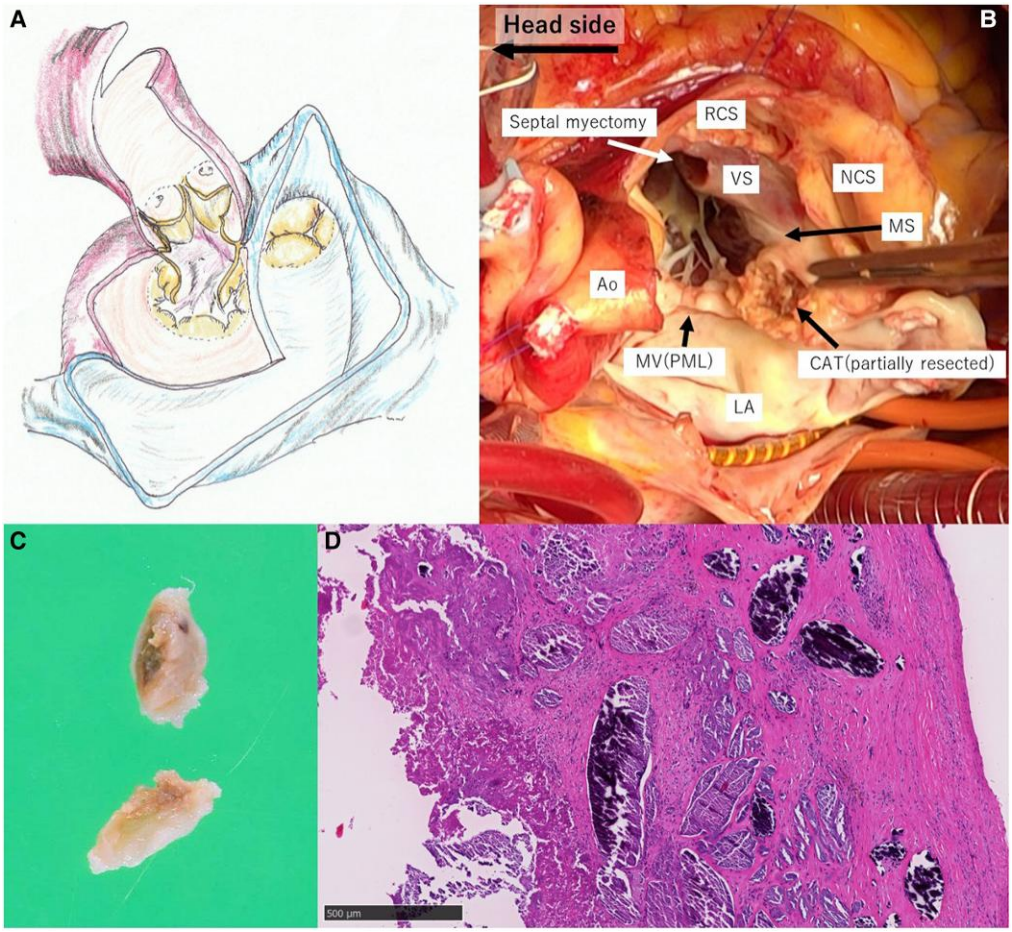


**Figure 4** (*A*) Illustration of incisional view. The transseptal atriotomy and aortotomy were performed to across the left and non-coronary commissure into the anterior mitral valve leaflet with opening the IVF. (*B*) The gross findings of aortic valve cusps during surgery display a calcified amorphous tumour and septal myocardium protruding into the left ventricular outflow tract. RCS, right coronary sinus; VS, ventricular septum; NCS, non-coronary sinus; MS, membranous septum; CAT, calcified amorphous tumour; LA, left atrium; MV, mitral valve; PML, post-mitral leaflet; Ao, aorta, (*C*) CAT specimen after resection. CAT was covered with fibrous tissue. (*D*) Histopathology image of CAT. Most of the unstructured deposits are calcification. Calcium deposits are present in the cavity along with eosinophilic deposits. Macrophage infiltration was observed in the background.

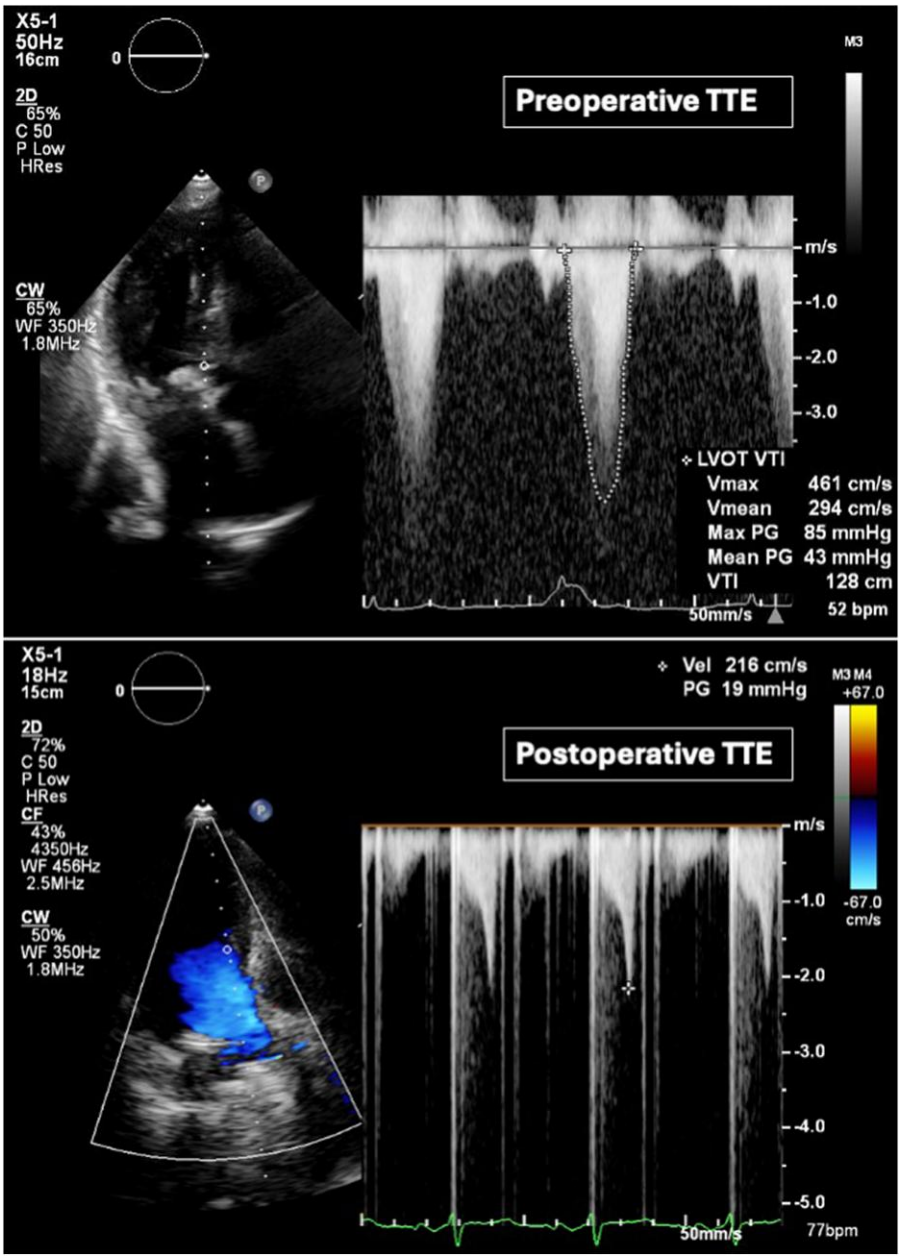


**Figure 5** Transthoracic echocardiogram. Preoperatively, the maximum pressure gradient in the left ventricular outflow tract was 85 mmHg with the valsalva manoeuvre, but it improved to 15 mmHg post-operatively.

replacement with Commando Operation, and myectomy, as calcification spread from IVF to AML and NCC. Resection of only the protruding portion of the CAT and the septal myocardium may increase the risk of embolization. This is because removing the outer membrane of CAT can result in the release of calcified debris, which may lead to embolization.

## Conclusion

Because CAT is a recognized disease in cardiac tumourigenesis, its accumulation of more CAT cases is important. Moreover, CAT may contain factors other than calcium and phosphate metabolism that contribute to its formation in patients with normal renal function.

## Lead author biography

Graduated from Kanazawa Medical University in 2014 and has been working as a cardiologist in the Department of Cardiology at Gifu University School of Medicine since 2016. Specialized in clinical and research imaging fields such as echocardiography and cardiac CT.

## Supplementary material

Supplementary material is available at *European Heart Journal – Case Reports* online.


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## Data availability

No new data were generated or analysed in support of this research.

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