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OVERCOMING CRISIS: INNOVATIVE APPROACH TO SEPTAL ABLATION IN HYPERTROPHIC CARDIOMYOPATHY WITH ISCHAEMIA

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ABSTRACT
Hypertrophic Cardiomyopathy (HCM) is a genetic disorder with an autosomal dominant mode of inheritance. 1:500 in the general population are affected by it. It can be generalised or restricted largely to the interventricular septum. Septal hypertrophy can lead to Left Ventricular Outflow Tract Obstruction (LVOTO), which occurs in 25% of cases resulting in poor prognosis. However, Ethyl Alcohol Ablation has been introduced as a method to reduce the LVOTO especially in patients refractory to medical therapy. The coexistence of HCM and Coronary Artery Disease(CAD) is rare. Here we present a case report of a 40-year-old lady, who presented with HCM and CAD, and had undergone Percutaneous Coronary Intervention(PCI) and Ethyl Alcohol Ablation due to unresolving symptoms.

KEYWORDS: *Hypertrophic Cardiomyopathy, Coronary Artery Disease (CAD), LVOTO, Ethyl Alcohol Ablation.*

INTRODUCTION

Hypertrophic Cardiomyopathy (HCM) is a genetic heart disease which is the most common cause of Sudden Cardiac Death (SCD) in young athletes and can lead to heart failure, atrial fibrillation, and ischaemic symptoms.[1] Asymmetric Septal Hypertrophy (ASH) is a subtype of HCM and is characterized by asymmetric thickening of the interventricular septum, which can lead to Left Ventricular Outflow Tract Obstruction (LVOTO) and Mitral Regurgitation (MR).[2] Adult patients with HCM can also develop atherosclerotic Coronary Artery Disease (CAD).[3,4] In cases of HCM, angina may be misleading and result in a failure to recognize coexisting CAD. Here, we report a rare case of a 40-year-old HCM patient with CAD. In this patient, we could successfully manage ischaemic symptoms using successive procedures of Percutaneous Coronary Intervention (PCI) and Ethyl Alcohol Ablation of the septal branches of the Left Anterior Descending (LAD) artery.

CASE REPORT

A 40-year-old lady was admitted with the complaint of severe compressive central chest pain at rest for one day which was non-radiating and was associated with shortness of breath. She mentioned that she has been suffering from exertional breathlessness for the last 3 years and symptoms have progressively worsened recently. She experienced four episodes of cardiac syncope while doing moderate physical activities in the span of 8 years. She was hypertensive and had bronchial asthma. On examination, she was dyspnoeic and was in a propped up position. Her Heart Rate was 110 bpm, Blood Pressure was 110/60 mmHg, Respiratory Rate was 26 breaths/min, afebrile and JVP was raised (8 cm from the sternal angle). On precordial examination, the apex beat was forceful and sustained. The First Heart Sound was soft and the Second Heart Sound was normal and a pansystolic murmur could be heard near the apex radiating to the axilla. There was also a non-radiating ejection systolic murmur at the left parasternal edge. On respiratory system examination, there was bilateral vesicular breath sound with prolonged expiration with diffused rhonchi and inspiratory crepitations in the lower zones.

Initial investigations revealed both samples of Troponin-I were negative. NT pro BNP was elevated. ECG was done revealing Left Atrial Enlargement (LAE) and poor R wave progression **(Figure 1)**.

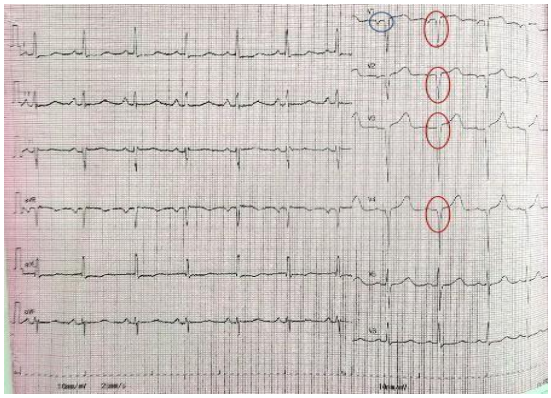

Figure 1: ECG showing a P- terminal force of >1 mm and poor progression of R wave.

Chest X-ray P/A view revealed cardiomegaly. An Echocardiography was done that unveiled a Hypertrophic Obstructive Cardiomyopathy (HOCM) with Systolic Anterior Motion of Anterior Mitral Leaflet with a severe LVOT gradient (132 mmHg) and a consequent moderate mitral regurgitation **(Figure 2 A,B)**.

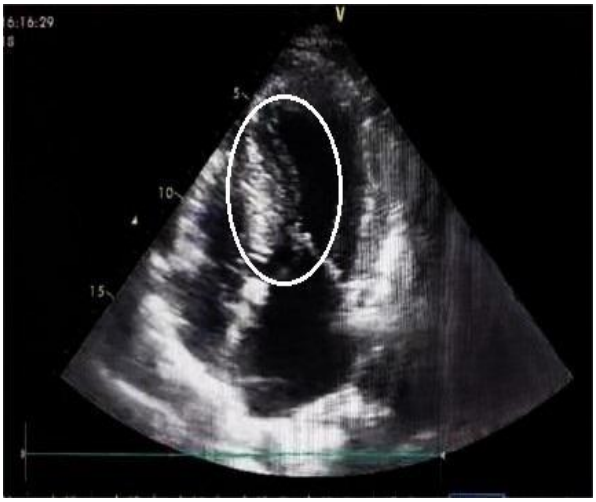

Figure 2: (A) Echocardiography revealing Septal Hypertrophy and systolic anterior motion of anterior mitral leaflet.

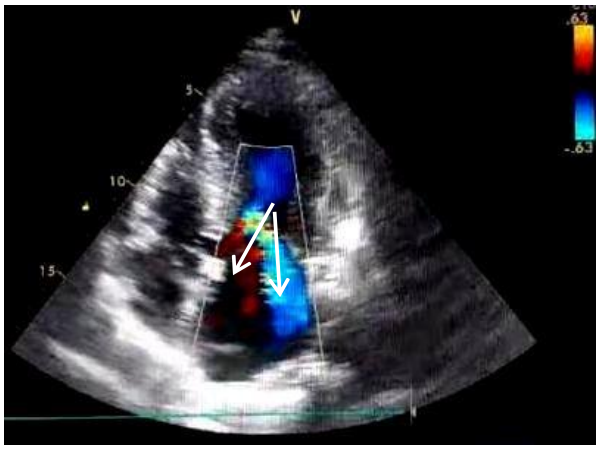

Figure 2: (B) Echocardiography revealing systolic anterior motion of anterior cusp of the mitral valve with non coaptation and Mitral Regurgitation (MR).

The patient was treated for Acute Heart Failure with intravenous Furosemide and oral Spironolactone and Ivabradine. Due to progressive worsening of symptoms, a Coronary Angiogram (CAG) was performed, discovering 90% stenosis in the Posterior Descending Artery (PDA) branch of Right Coronary Artery (RCA) during diastole and 40-50% stenosis in proximal Left Anterior Descending (LAD) with a myocardial bridge in mid LAD. Our plan was to do 2 procedures at an interval of 4-6 weeks: Percutaneous Coronary Intervention (PCI) to the PDA and septal ablation with ethyl alcohol. Considering recent worsening of symptoms, we first did PCI to PDA and a 2.5 X 23mm Drug Eluting Stent (DES) was deployed **(Figure 3A,B).**

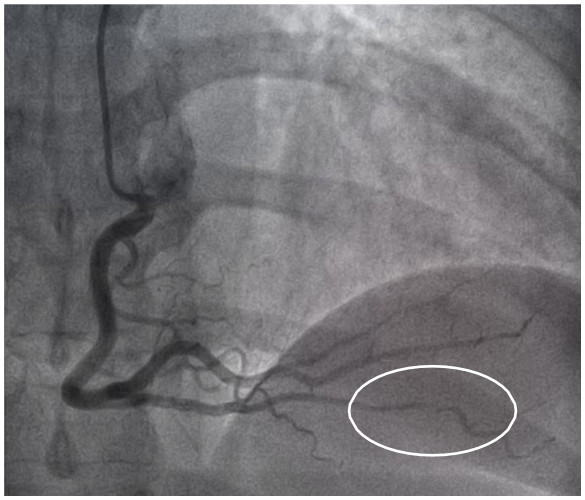

Figure 3(A): CAG revealing 90% occluded Posterior Descending Artery.

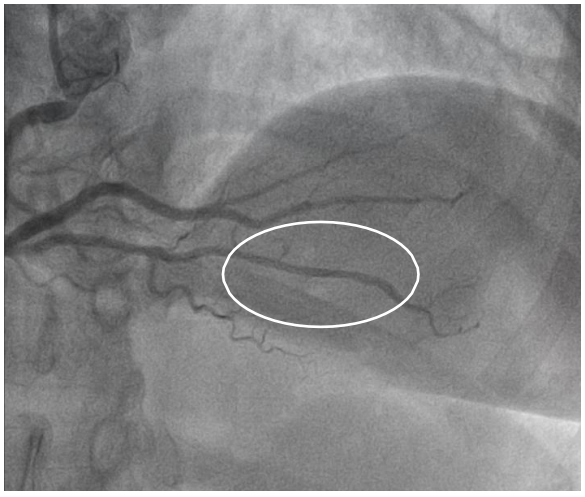

Figure 3(B): Result of PCI to PDA.

Post PCI, along with heart failure medication, the patient was on Prasugrel 10 mg, Aspirin 75 mg, Rosuvastatin 10 mg and Glyceryl Trinitrate (GTN) 2.6 mg. She gradually improved and was discharged 4 days after PCI.

Unfortunately, she was readmitted 14 days later with severe angina at rest and breathlessness. A check CAG revealed a patent stent in PDA and a decision for urgent

Alcohol Septal Ablation was made. But we had few challenges. We did not have the conventional Over The Wire (OTW) balloon (due to post pandemic crisis of hardware and no vendor could supply it urgently) and the patient was in acute heart failure. A Heart team discussion suggested that the OTW balloon principle of "block and delivery" could also be applied by simultaneous use of balloon and microcatheter. After discussing the pros and cons of alternative method as well as the surgical myomectomy option, the patient and her relatives decided to go on with the proposed alternative available treatment.

Temporary pacing was inserted through femoral vein. A 5Fr Pigtail catheter was inserted through the femoral artery and LVOT gradient was measured which was 108 mmHg. A JL 6Fr guiding catheter was used to engage the Left Coronary Artery (LCA). Angiogram was done. The largest septal branch was the second septal artery which is responsible for the irrigation of the mid-basal portion of the interventricular septum, as seen after echocardiographic contrast injection **(Figure4).**

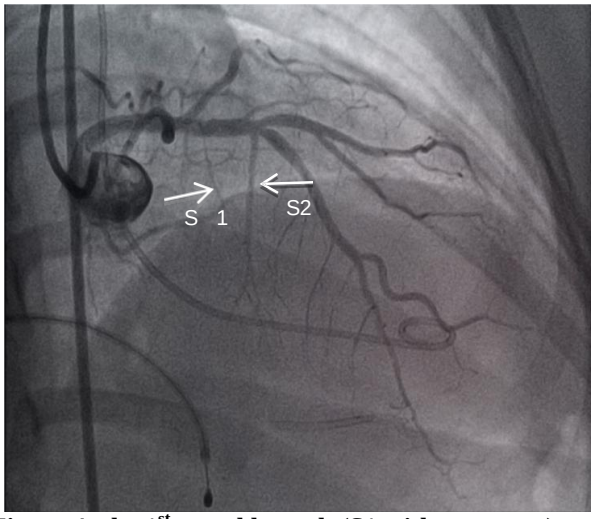

Figure 4: the 1st septal branch (S1 with an arrow) and 2nd septal branch (S2 with an arrow).

Over a 0.014" 180 cm Runthrough wire, a micro- guide catheter (Fine Cross Terumo) was passed into the septal branch. The wire was removed and then re-introduced into the septal branch alongside the micro-guide catheter. A 2.0 X 12 mm non compliant balloon was passed into the septal artery, keeping the distal end of the balloon 5-10 mm proximal to the micro-guide catheter tip. The balloon was inflated at 12 ATM and angiography was performed in 2 ways: first, contrast is injected from the guide catheter to ensure no flow into the occluded septal artery, and second, selective angiography through the micro-guide catheter lumen was performed to ensure no reflux of contrast into the LAD or contrast traveling into collateral arteries. Then, keeping the balloon inflated, 2ml of 95% Ethyl Alcohol was infused slowly over 2 minutes through the micro-guide catheter followed by 0.3ml normal saline flush. The balloon was kept inflated for 10 minutes **(Figure 5)**.

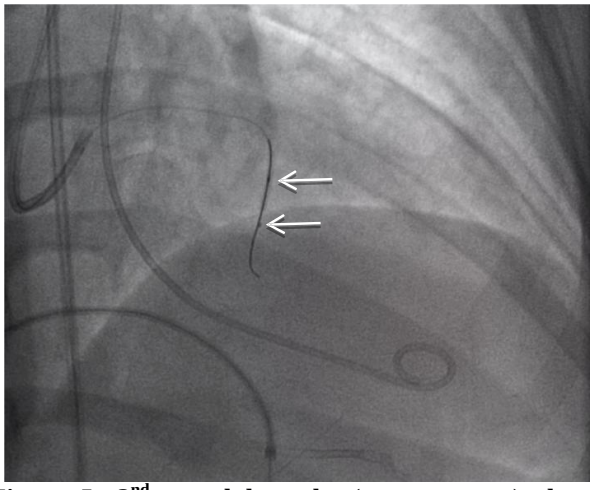

Figure 5: 2nd septal branch: (upper arrow) shows inflated balloon and (lower arrow) shows tip of the microcatheter.

Then both the micro-guide catheter and the balloon were removed. Angiogram showed complete blockage of the distal part of target septal branch. There was slow flow (TIMI-2) in LAD which improved after intra coronary injection of Glyceryl Trinitrate (GTN). However, after blocking the second septal artery, there was no improvement in LVOT gradient. So we searched for another septal branch with therapeutic potential. Despite the reduced caliber and length, catheterization of the 1st septal branch with balloon-catheter occlusion resulted in LVOT gradient decrease which was restored to the baseline after balloon deflation.

Moreover, the injection of echographic contrast revealed that the 1st septal branch had a larger irrigation area of the mid-basal portion of the interventricular septum, when compared to the 2nd. Based on these findings, we performed alcoholization of the 1st septal branch by the same technique. **(Figure 6A)**

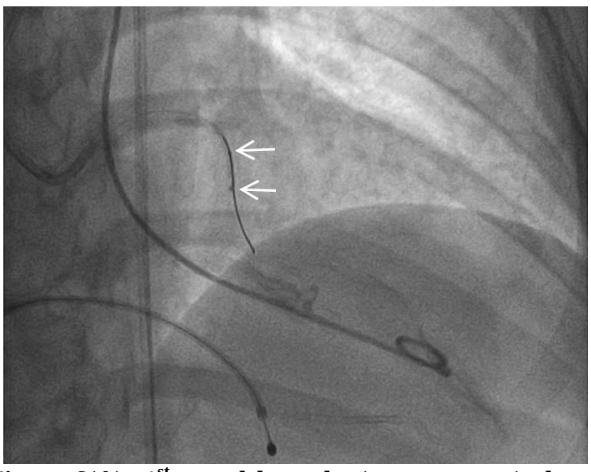

Figure 6(A): 1st septal branch: (upper arrow) shows inflated balloon and (lower arrow) shows tip of the microcatheter.

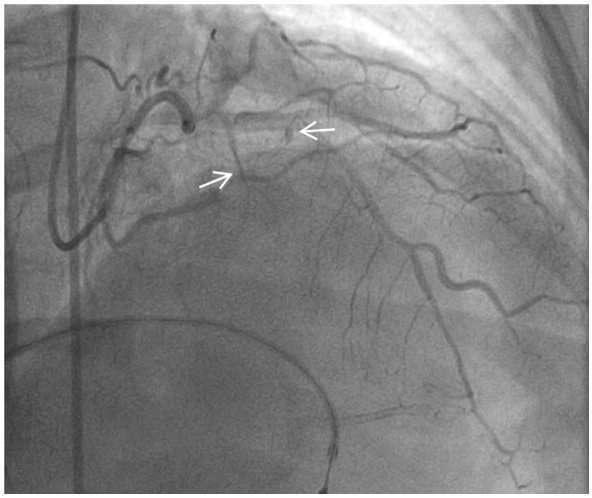

Figure 6(B): Both septal branches totally occluded after septal ablation (indicated with arrows)

Haemodynamic assessment revealed the LVOT gradient came down to 17 mmHg from 108 mmHg **(Figure7A,B)**.

Figure 7(A): LVOT gradient before septal ablation (108 mmHg).

Figure 7(B): LVOT gradient after septal ablation (17 mmHg).

Bedside transthoracic echocardiogram revealed normal LVOT gradient **(Figure8A,B).**

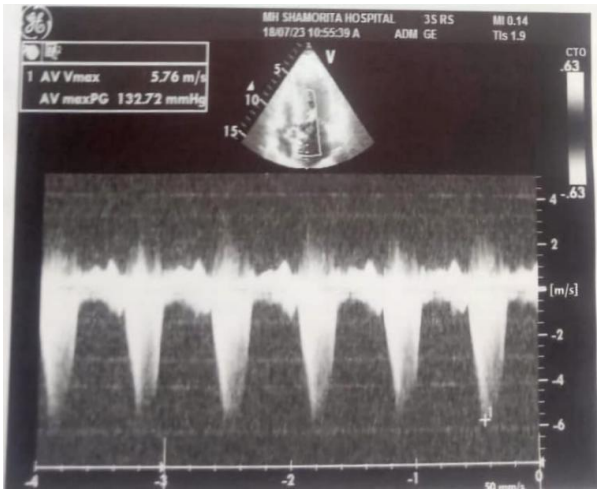

Figure 8(A): ECHO (2D) LVOT gradient before septal ablation.

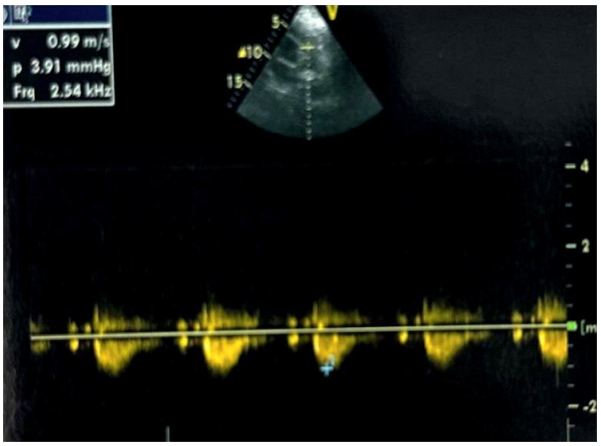

Figure 8(B): ECHO (2D) LVOT gradient after septal ablation.

The patient remained haemodynamically stable throughout the procedure. Narcotic analgesic was used during the procedure. After the procedure, the temporary pacemaker was kept and as there was no AV block for 48 hours, it was then removed. The patient made a good recovery and was discharged 5 days after the procedure. We did a follow up after one year and the patient did not have any symptoms. Her LVOT gradient on echocardiogram was normal.

DISCUSSION

Approximately 25% of HCM patients have evidence 5 of ischemia during regular daily activites. The presence of ischemia has been associated with a 6 worse prognosis in HCM. The management of patients with ASH and ischaemic symptoms is challenging, and several treatment options have been proposed, including medical therapy, septal reduction therapy, and dual-chamber pacing.[7]

HCM coexisting with CAD was originally de- scribed in 1972 and is extremely rare. Timely CAG is the gold standard for diagnosing HCM comorbid with CAD. Revascularization is, however, performed in only a

small number of patients with HCM. Our young patient's clinical symptoms were initially alleviated after blood flow was improved in her RCA. But as the acute heart failure symptoms reappeared despite optimum tolerated medications, we had to consider further interventions on an urgent basis.

ESC Guidelines on diagnosis and management of hypertrophic cardiomyopathy recommend an invasive treatment to reduce LVOTO in patients with an LVOT gradient ≥ 50 mmHg, moderate to severe symptoms [New York Heart Association (NYHA) functional Class III–IV] and/or recurrent exertional syncope in spite of maximally tolerated drug therapy.[8]

Ethyl Alcohol Ablation leads to localized myocardial necrosis and thinning of the hypertrophied septum. It has been shown to be effective in reducing LVOTO and improving symptoms in patients with HCM and refractory heart failure or ischaemic symptoms.[7]

On the other hand, surgical septal myectomy has been shown to improve symptoms and survival in patients with obstructive HCM.[9] Several meta- analyses have shown that both surgery and septal alcohol ablation improve functional status with a similar procedural mortality.[10,11] However, decision to undergo surgery should be made on a case-by-case basis after careful consideration of the patient's overall health and individual needs. Our patient had acute heart failure and she refused the surgical option.

Another challenge was the unavailability of OTW balloon. So, we had to think of an alternative. The 'block and deliver' technique[12,13,14]: simultaneous delivery of an occluding balloon and a microcatheter through a single guide catheter has been described in managing coronary perforation in distal vessel. In this technique, coil advancement in the distal lumen is possible even in the presence of an inflated balloon on top of the microcatheter. In addition, selective tip injections through the microcatheter allow assessment of hemostasis without the need to repeatedly deflate the occlusive balloon. In our patient, we used the same principle: The inflated balloon was used to prevent any retrograde leakage of Alcohol (Block) and the microcatheter was used to inject alcohol in the distal septal branch (Deliver).

CONCLUSION

This case report emphasizes the utilization of available materials in managing acute situation even in the scarcity of ideal resources. However, it also highlights the complexity of managing patients with multiple cardiac issues and the role of ethyl alcohol ablation as an effective, minimally invasive intervention for symptomatic patients with ASH.

I. Informed Consent

The authors confirm that written consent for submission and publication of this case report including image(s) and associated text has been obtained from the patient.

II. Potential Conflict of Interest

No potential conflict of interest relevant to this article was reported.

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