Cardiology and Angiology: An International Journal

Volume 13, Issue 4, Page 54-63, 2024; Article no.CA.117267
ISSN: 2347-520X, NLM ID: 101658392

A Tale of Grumpy Neighbours: Angina from Left Main Coronary Artery Compression in Severe Pulmonary Hypertension with Large ASD

Kalyan S Munde [a++], Anagh T S [a#*], Samkit Mutha [a†], Jayakrishna Niari [a†], Mohan Paliwal [a#] and Hariom Kolapkar [a#]

[a] Department of Cardiology, Grant Govt. Medical College and Sir JJ Group of Hospitals Mumbai, India.

Authors' contributions

This work was carried out in collaboration among all authors. All authors read and approved the final manuscript.

Article Information

DOI: https://doi.org/10.9734/ca/2024/v13i4440

Open Peer Review History:
This journal follows the Advanced Open Peer Review policy. Identity of the Reviewers, Editor(s) and additional Reviewers, peer review comments, different versions of the manuscript, comments of the editors, etc are available here: https://www.sdiarticle5.com/review-history/117267

Case Report

Received: 10/03/2024
Accepted: 16/05/2024
Published: 28/09/2024

ABSTRACT

This report presents a case of a 42-year-old patient diagnosed with ostium secundum atrial septal defect (ASD) with severe pulmonary arterial hypertension (PAH). The patient presented with progressive chest pain and dyspnea and was found to have a lesion in the left main coronary artery (LMCA) ostium by coronary angiography. Computed tomography (CT) imaging revealed a significant enlargement of the main pulmonary artery (MPA), which indicated possible compression

[++] HOD;
[#] Senior Resident;
[†] Assistant Professor;
**Corresponding author: E-mail: anaghtshetru@gmail.com;*

Cite as: *Munde, Kalyan S, Anagh T S, Samkit Mutha, Jayakrishna Niari, Mohan Paliwal, and Hariom Kolapkar. 2024. "A Tale of Grumpy Neighbours: Angina from Left Main Coronary Artery Compression in Severe Pulmonary Hypertension With Large ASD". Cardiology and Angiology: An International Journal 13 (4):54-63. https://doi.org/10.9734/ca/2024/v13i4440.*

of the LMCA by the pulmonary artery which was subsequently confirmed by coronary angiography and intravascular ultrasound (IVUS). Our patient underwent IVUS guided percutaneous coronary intervention by which a stent was deployed in the ostial LMCA, leading to near resolution of the patient's symptoms. Currently, the patient is under follow-up. This case highlights an often-overlooked cause of chest pain and dyspnea in patients with PAH. Although these patients often experience typical and atypical angina due to elevated right-sided pressures, the current report reveals that external compression of the LMCA by an enlarged pulomonarya artery(PA) can also cause coronary ischemia. Physicians should consider LMCA compression when assessing PAH patients with chest pain, as percutaneous coronary intervention and stenting can safely and effectively manage this condition.

Keywords: Left main coronary artery; main pulmonary artery; pulmonary hypertension; atrial septal defect; coronary angiography; intravascular ultrasound; computed tomography coronary angiography.

1. INTRODUCTION

“Pulmonary arterial hypertension entails a poor prognosis irrespective of the underlying aetiology” [1,2,3]. “Typical symptoms are largely nonspecific, including dyspnea, fatigue, and chest pain on exertion. Angina-like symptoms are frequent despite angiographically normal coronary arteries, and classically, have been attributed to right ventricular ischemia resulting from the increased metabolic needs of the overloaded, hypertrophied right ventricle or painful distention of the PA” [1,2,3].

“A severely dilated PA may compress adjacent anatomical structures including the LMCA (causing myocardial ischemia), the left recurrent laryngeal nerve (causing hoarseness; i.e., Ortner's syndrome), and the tracheobronchial tree (large airway obstruction)” [4].

“External compression of the LMCA by a dilated MPA is a rare condition, but it can cause chest pain in patients with pulmonary hypertension. However, extrinsic compression of the LMCA by a dilated pulmonary artery main trunk is increasingly recognized as a cause of angina in PAH. Despite multiple case reportsthe incidence of LMCA compression in PAH is not well established” [5,6] “A small case series reported that, of 26 patients with PAH and angina, 7 (26.9%) had LMCA compression” [7].

Along with angina, LMCA compression may be associated with additional complications of severe myocardial ischemia, including myocardial infarction (MI) [8], left ventricle dysfunction, arrhythmia, and eventually, sudden death[1] Because >25% of deaths in patients with PAH are related to sudden death [1], some of these events might be attributable to LMCA compression, a potentially correctable complication.

“Currently, computed tomography coronary angiography (CTCA) provides a precise assessment of the anatomical relationship between the LMCA and the enlarged PA” [7].

Traditionally, coronary artery bypass graft surgery is used to treat LMCA disease. However, in the case of LMCA compression caused by a dilated PA, coronary angioplasty with stenting has shown promising results and might be a better option for some high-risk patients who cannot undergo surgery.

2. CASE PRESENTATION

A 42-year-old male patient, with no medical history in the past, visited us with progressively worsening dyspnea New york heart association (NYHA) class II and exertional chest pain over a period of two years. On physical examination, we observed pectus excavatum of the anterior chest wall (Fig. 1), a blood pressure of 100/80 mmHg, a heart rate of 80 beats/minute, and no signs of left or right-sided heart failure. The saturation levels in all four limbs (Table 1) were checked and recorded at room temperature.

On cardiac examination, the patient was found to have a loud P2 with an ejection systolic murmur of grade 3/6 in the pulmonary area. However, there was no elevated jugular venous pressure or pedal edema observed. The patient's breath sounds were normal. The ECG indicated right ventricular hypertrophy with a strain pattern (Fig. 2), while the chest X-ray showed a prominent pulmonary artery shadow, suggesting a dilated pulmonary artery (Fig. 3). During a 6-minute walk test, the patient experienced chest pain around

the chest after walking for 4 min and was unable to complete the test. The distance covered by the patient was 280m and pulmonary function testing was suggestive of mild restrictive pattern.

Further testing with transthoracic echocardiography showed a large Ostium Secundum atrial septal defect (OS-ASD) of size 20.3mm with a bi-directional shunt (Figs. 4,5,6,7). The right atrium (RA) and right ventricle (RV) were both dilated, with an estimated systolic RV pressure (RVSP) of 107 mmHg and a tricuspid annular plane systolic excursion (TAPSE) of 15.5 mm. The left ventricle (LV) appeared 'D'-shaped and small, with an end-diastolic diameter of 18.9 cm. However, the LV systolic function was normal, with an ejection fraction of 62%. The main pulmonary artery was also dilated, with a diameter of 32 mm. After performing a coronary angiogrpahy from the right femoral artery, a short segment eccentric ostial LMCA stenosis was discovered, followed by a mid left anterior descending artery (LAD) myocardial bridge (Fig. 8). There were no other atherosclerotic changes or stenosis of the coronary arteries. IVUS showed that the ostium of the LMCA was narrowed to a slit without evidence of underlying atherosclerosis (Fig. 9). Cardiac computed tomography (ECG-gated multidetector computed tomography [MDCT]) confirmed high origin of LMCA from the left cusp with significant narrowing at its origin and no obvious wall calcification (CADRADS 3/4) caused by a severely dilated PA trunk (Figs. 10,11). A large OS- ASD of 22mm with dilated RA, RV, and PA was also confirmed. The aortopulmonary ratio was 0.7, which was suggestive of pulmonary hypertension.

Table 1. Saturation of all limbs

R UL = 91%	L UL = 91%
R LL = 92%	L LL = 91%

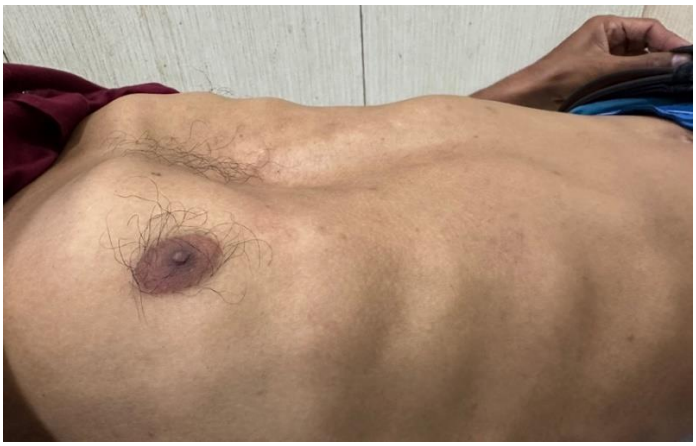

Fig. 1. Pectus excavatum

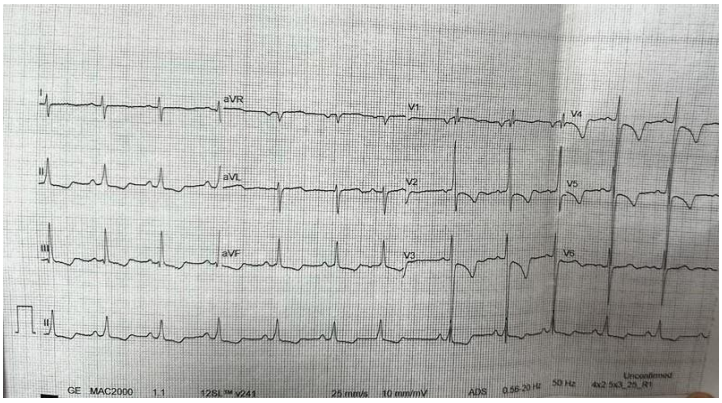

Fig. 2. Right ventricular hypertrophy with strain pattern

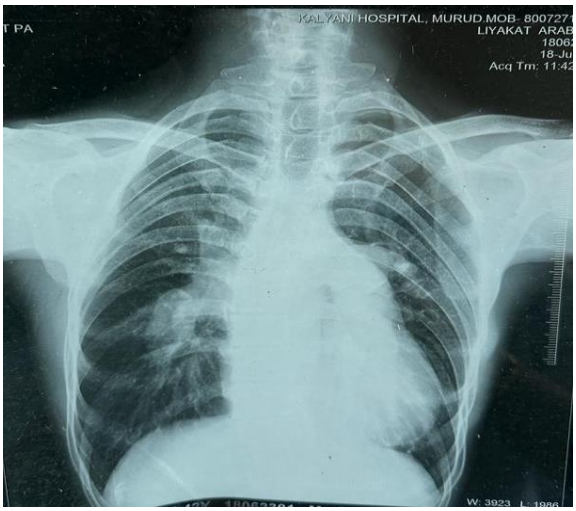

Fig. 3. CXR showing Prominent pulmonary artery

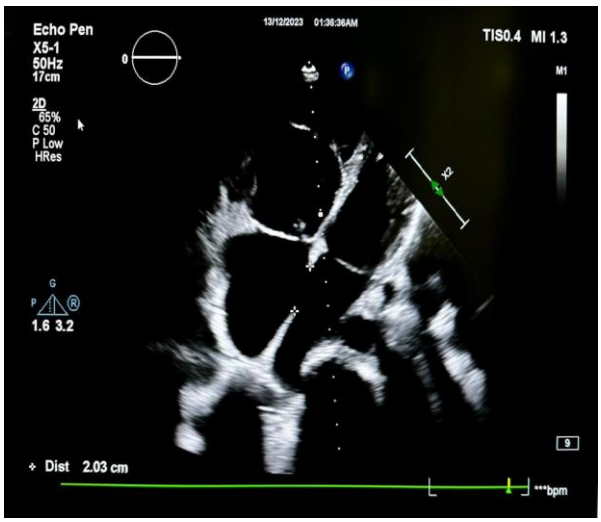

Fig. 4. Ostium secundum atrial septal defect of size 20.3mm

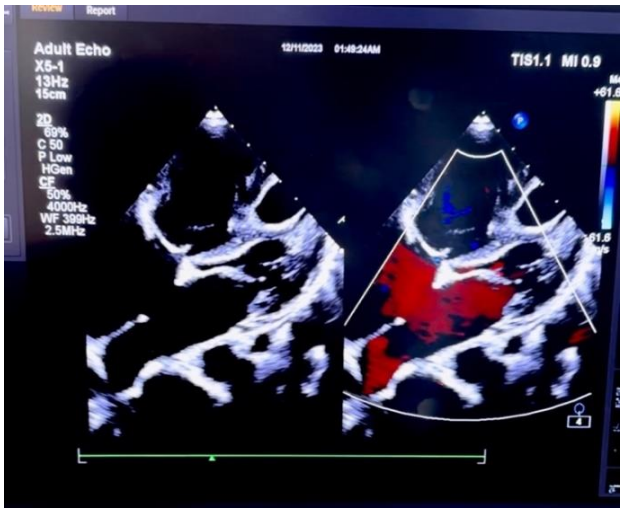

Fig. 5. Colour doppler across septal defect

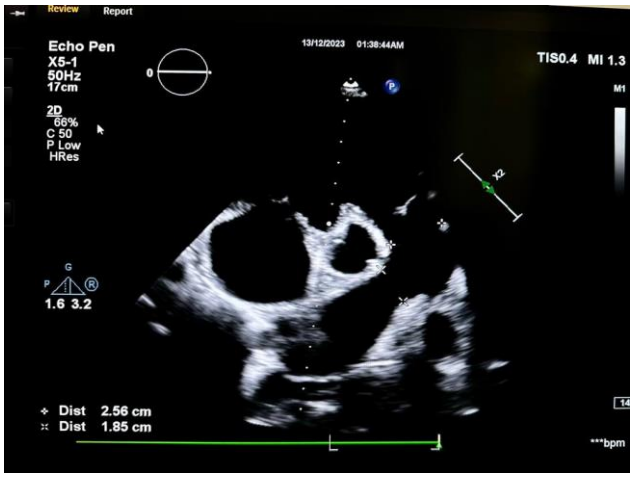

Fig. 6. Dilated pulmonary artery

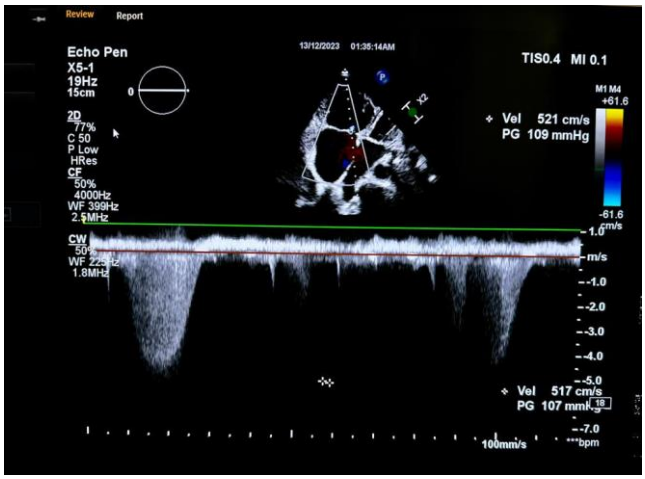

Fig. 7. Right ventricular systolic pressure – 107mmhg

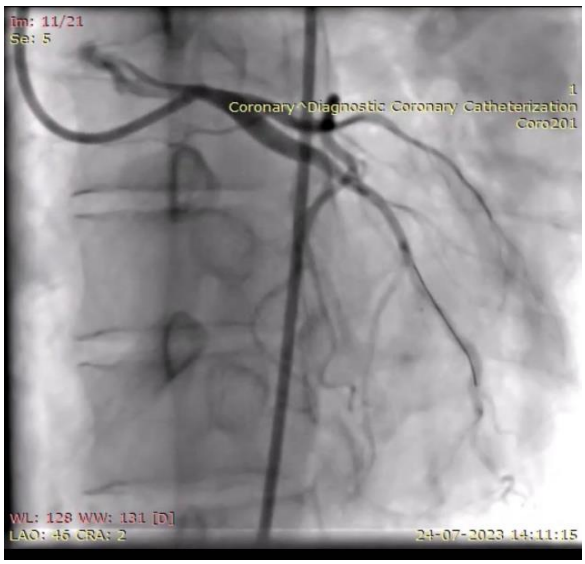

Fig. 8. Coronary angiography demonstrating eccentric 60% stenosis of the ostium of the left main coronary artery (LMCA). No other atherosclerotic changes noted

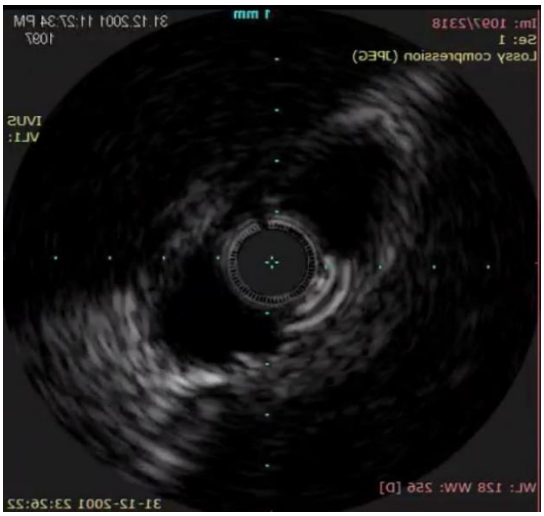

Fig. 9. IVUS showing ostial LMCA narrowed to a slit

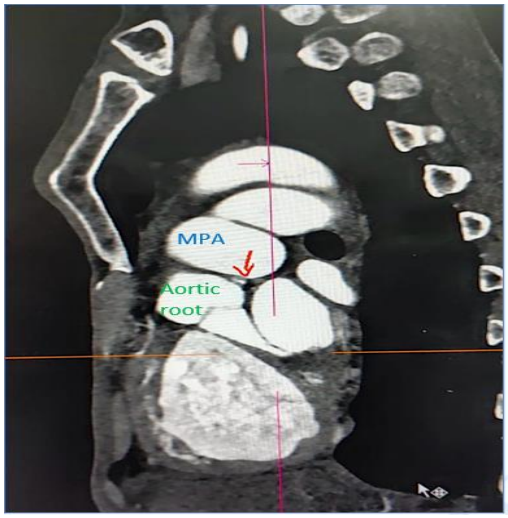

Fig. 10. High origin of LMCA from left cusp with significant narrowing at its origin and no obvious wall calcification (CADRADS 3/4)

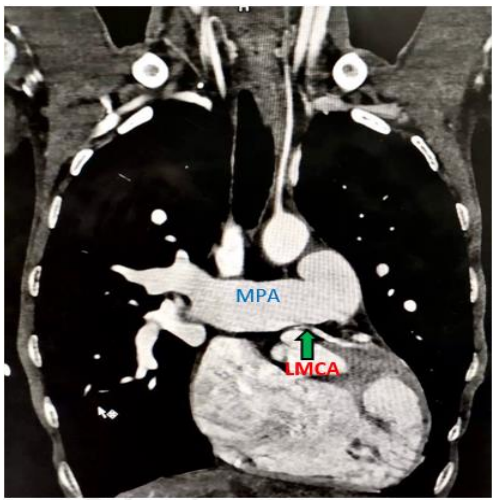

Fig. 11. External compression of LMCA at the ostium by dilated PA

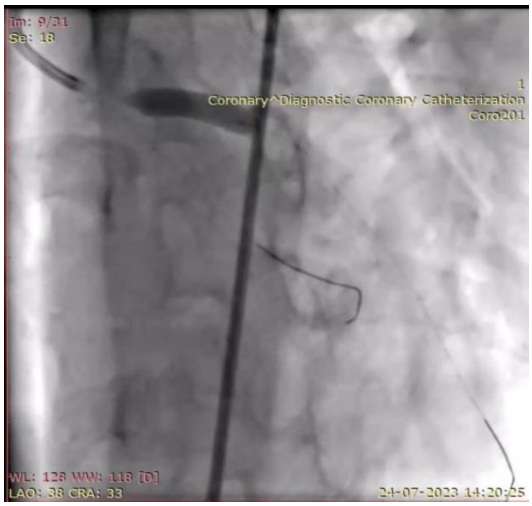

Fig. 12. Left main coronary artery from the ostium stented using RENOFIT 6x15mm bare metallic stent

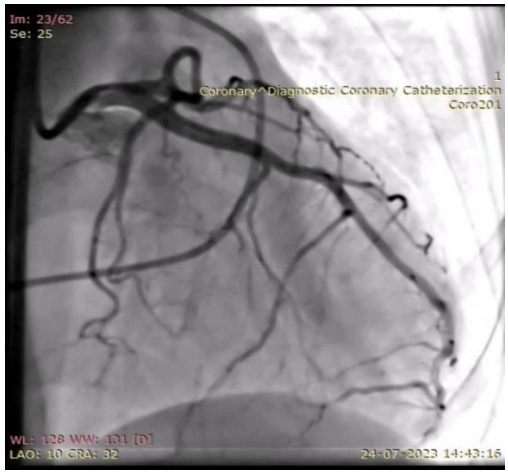

Fig. 13. Stent well opposed, TIMI 3 flow

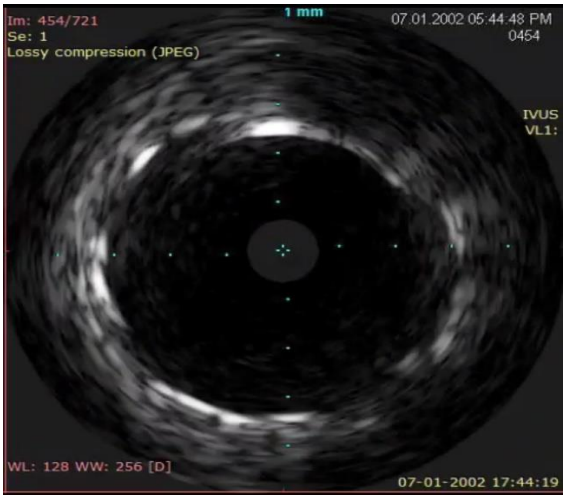

Fig. 14. IVUS of LMCA post stent deployment showing well opposed stent with large calibre vessel. (MSA – 9.4mm2)

Table 2. LVEDP post balloon occlusion

Pre occlusion	3 min	6 min	10 min
LVEDP (mmhg) – 5	16	30	37

Table 3. Pre-procedure and post-procedure assessment

	Pre procedure	Post procedure (3 months)
Symptoms	Angina +	No angina
	Dyspnea NYHA III	NYHA I
6MWD	280	430m
RVSP	107 mmhg	86mmhg

2.1 Procedure

Our patient underwent a percutaneous coronary intervention (PCI) of the left main artery ostium using a bare metallic stent RENOFIT 6*15mm guided by IVUS. (Figs. 12,13,14). The post-procedure stent was well opposed, and the narrowing of the LMCA ostial disappeared. There was no evidence of flap, dissection or residual ostial lesion post stenting. The same findings were confirmed by IVUS and the minimal stent area (MSA) was found to be 9.4 mm^2. (Fig. 14). To assess the reversibility of PAH, cardiac catheterization was done, which showed irreversible PAH (PVR 11 wood units). An attempt to close the atrial septal defect (ASD) with a device was not made in this case due to increased pulmonary vascular resistance and a significant increase in LVEDP on balloon occlusion of the septal defect (Table 2). The patient's symptoms have been resolved after the PCI, and he is currently undergoing medical management for pulmonary hypertension and is on follow-up.

3. DISCUSSION

“Extrinsic compression of the LMCA due to a dilated PA has been previously reported” [2]. “This condition can present with exercise-related chest pain, cardiogenic shock, malignant arrhythmias caused by myocardial ischemia, or sudden death. Left coronary artery compression syndrome was first described by Corday et al., in 1957 as compression of the LMCA between the aorta and an enlarged main PA” [9]. “It is usually seen with a congenital cardiac defect, most commonly an atrial septal defect, ventricular septal defect, or tetralogy of Fallot” [10].

“The incidence of LMCA compression due to PA dilatation is not well established, but it ranges between 5% and 44% according to different case series” [7,11].

“Because of a low pretest probability for atherosclerotic coronary artery disease in young PH patients, coronary angiography is rarely performed. However, the extrinsic compression of the LMCA due to an enlarged PA is becoming increasingly recognizable and should always be considered in PH patients with exercise-induced chest pain, which is the most common clinical presentation of this problem” [12,13]. “In one study, the origin of the LMCA from the right sinus of Valsalva was considered to convey a higher risk for extrinsic compression compared with the normal origin from the left sinus” [10]. “In another study, risk factors predisposing to LMCA compression in PAH were younger age, severe pulmonary trunk dilatation (>40 mm; normal: 25–30 mm), and a PA trunk/aorta ratio >1.2 (normal: 1.0)” [7].

Our patient was young and suffered from severe pulmonary hypertension. The pulmonary trunk size was 35mm with a PA trunk/aorta ratio of 1.45, with MPA 35mm and aorta 24mm. As there were no signs of atherosclerosis, a short bare metallic stent was used, and aggressive post dilation was avoided. Additionally, as left main artery stenting was involved, IVUS was utilized for stent optimization to avoid any complications [14].

In cases where extrinsic compression of the LMCA by an enlarged PA occurs, PCI is a reasonable first therapeutic option considering its high likelihood of success. The procedure provided persistent symptomatic and prognostic benefits for our patient [15].

The patient underwent IVUS-guided PCI for the left main ostium and was discharged on dual antiplatelet therapy, tadalafil and ambrisentan on

discharge. On follow-up up patient has been symptomatically better and his exercise capacity has improved

4. CONCLUSION

Extrinsic compression of the LMCA by an enlarged PA is rare but a significant cause of angina in patients with PAH. This condition should be suspected in a case of PAH where the patient shows typical or atypical anginal symptoms. The most appropriate initial test to evaluate this condition is CTCA. Coronary angiography is required to confirm LMCA stenosis and to enable PCI with stent deployment. PCI with stent deployment is a safe and effective treatment that can lead to long-term improvement in patients' symptoms.

Physicians should consider LMCA compression when assessing PAH patients with chest pain, as percutaneous coronary intervention and stenting can safely and effectively manage this condition.

In summary, extrinsic compression of the LMCA by an enlarged PA can cause chest pain in patients with advanced PAH, and CTCA is the first step in evaluating it and PCI with stent deployment is a safe and effective treatment for this condition.

DISCLAIMER (ARTIFICIAL INTELLIGENCE)

Author(s) hereby declare that NO generative AI technologies such as Large Language Models (ChatGPT, COPILOT, etc) and text-to-image generators have been used during writing or editing of this manuscript.

CONSENT

As per international standards or university standards, patient(s) written consent has been collected and preserved by the author(s).

ETHICAL APPROVAL

As per international standards or university standards written ethical approval has been collected and preserved by the author(s).

COMPETING INTERESTS

Authors have declared that no competing interests exist.

REFERENCES

1. Demerouiti EA. Manginas AN, Atha GD, Karatasakis GT. Complications leading to sudden cardiac death in pulmonary arterial hypertension. Respir Care. 2013;58(7):1246-54.
   DOI: 10.4187/respcare.02252.
2. MS. Lee J, Oyama R, Bhatia YH, Kim SJ. Park Left main coronary artery compression from pulmonary enlargement due to pulmonary hypertension: A contemporary review and arguments for percutaneous revascularization. Catheter Cardiovasc Interv. 2010;76(4) :543-50.
   DOI: 10.1002/ccd.22592.
3. Galiè N, Humbert M, Vachiery JL. *et al.* 2015 ESC/ERS guidelines for the diagnosis and treatment of pulmonary hypertension: The Joint Task Force for the Diagnosis and Treatment of Pulmonary Hypertension of the European Society of Cardiology (ESC) and the European Respiratory Society (ERS). Eur Heart J. 2016;37(1):67-119.
   DOI: 10.1093/eurheartj/ehv317.Epub 2015 Aug 29.
4. Dakkak W, Tonelli AR. Compression of adjacent anatomical structures by pulmonary artery dilation. Postgrad Med. 2016;128(5):451–459.
5. VA. de Jesus Perez F, Haddad RH, Vagelos W, Fearon J, Feinstein RT, Zamanian Angina associated with left main coronary artery compression in pulmonary hypertension. J Heart Lung Transplant. 2009;28(5):527-30.
6. Karrowni W, Sigurdsson G, PA. Horwitz Left main coronary artery compression by an enlarged pulmonary artery. JACC Cardiovasc Interv. 2013;6(1):e3-4.
   DOI: 10.1016/j.jcin.2012.09.010.
7. Mesquita SM, Castro CR, Ikari NM, SA, Oliveira AA, Lopes Likelihood of left main coronary artery compression based on pulmonary trunk diameter in patients with pulmonary hypertension. Am J Med.2004;116(6):369-74.
   DOI: 10.1016/j.amjmed.2003.11.015.
8. Vaseghi M, Lee JS, Currier JW. Acute myocardial infarction secondary to left main coronary artery compression by pulmonary artery aneurysm in pulmonary arterial hypertension. J Invasive Cardiol. 2007;19(12):E375-7.

9. Corday E, Gold H, Kaplan L. Coronary artery compression; an explanation for the cause of coronary insufficiency in pulmonary hypertension Trans Am Coll Cardiol. 1957;7:93-103
10. Kajita LJ, Martinez EE, Ambrose JA. *et al.* Extrinsic compression of the left main coronary artery by a dilated pulmonary artery: clinical, angiographic, and hemodynamic determinants Cathet Cardiovasc Interv. 2001;52(1):49-54.
11. Lee MS, Oyama J, Bhatia R, Kim YH, Park SJ. Left main coronary artery compression from pulmonary artery enlargement due to pulmonary hypertension: a contemporary review and argument for percutaneous revascularization. Catheter Cardiovasc Interventions 2010;76:543–550.
12. Kanjwal MY, Carlson DE Jr., Schwartz JS. Chronic/subacute total occlusion of the left main coronary artery—a case report and review of literature. Angiology 1999;50: 937–945.
13. Shen AY, Jandhyala R, Ruel C, Lundstrom RJ, Jorgensen MB. Predictors of survival after coronary bypass grafting in patients with total occlusion of the left main coronary artery. Am J Cardiol 1998;81: 343–346.
14. D'Alonzo GE, Barst RJ, Ayres SM. *et al.* Survival in patients with primary pulmonary hypertension. Results from a national prospective registry. ann intern med 1991; 115(5):343-9.
DOI: 10.7326/0003-4819-115-5-343.
15. Kajita LJ, Martinez EE, Ambrose JA, Lemos PA, Esteves A, Nogueira da Gama M, Jatene AD, Ramires JA. Extrinsic compression of the left main coronary artery by a dilated pulmonary artery: clinical, angiographic, and hemodynamic determinants. Catheter Cardiovasc Interventions 2001;52:49–54.

Peer-review history:
The peer review history for this paper can be accessed here:
https://www.sdiarticle5.com/review-history/117267