AUCTORES
Globalize your Research

Journal of Clinical Cardiology and Cardiovascular Interventions
Varshit Hathi *

Open Access

Case Report

A Case of Extrinsic Compression of the Left Main Coronary Artery by Dilated Pulmonary Artery in Eisenmengers Syndrome

Nagamani Alur Chikkabasavaiah, Beeresh Puttegowda, Yeriswamy, Ramesh Basavappa, Varshit Hathi*, Manjunath Cholenally Nanjappa,
Department of Cardiology, Sri Jayadeva Institute of Cardiovascular Sciences and Research

***Corresponding Author:** Varshit Hathi, Department of Cardiology, Sri Jayadeva Institute of Cardiovascular Sciences and Research.

Received Date: November 04, 2020; **Accepted Date:** December 23, 2020; **Published Date:** January 02, 2021

Citation: Nagamani A. Chikkabasavaiah., Beeresh Puttegowda, Yeriswamy, Ramesh Basavappa, Varshit Hathi, Manjunath C. Nanjappa,., (2020) A Case of Extrinsic Compression of the Left Main Coronary Artery by Dilated Pulmonary Artery in Eisenmengers Syndrome. *J. Clinical Cardiology and Cardiovascular Interventions,* 4(1); Doi:10.31579/2641-0419/104

Abstract:

The extrinsic compression of the left main coronary artery secondary to pulmonary artery trunk dilatation is a relatively rare syndrome. [1,2] It is mostly associated with congenital acyanotic heart disease, idiopathic pulmonary arterial dilatation or primary pulmonary hypertension. [3,4] Most cases of pulmonary artery hypertension with no atherosclerotic risk factors rarely undergo coronary angiography, and hence, diagnoses are seldom made and proper management is delayed.We describe a patient with Ostium secundum atrial septal defect with severe pulmonary hypertension who presented with clinical angina ,diagnosed to have left main coronary artery compression by CT angiography ,underwent successful percutaneous coronary intervention with good symptomatic relief.

Key words: pulmonary artery dilatation; left main coronary artery

Introduction:

Angina- like chest pain is a common symptom in patients with pulmonary hypertension.It has been proposed that the pain may be due to right ventricular ischemia or pulmonary artery dilatation. [5,6] The extrinsic compression of the left main coronary artery secondary to pulmonary artery dilatation is another potential mechanism of chest pain. It is a rare syndrome that has been associated with severe pulmonary hypertension and sometimes associated with angina ,left ventricular ischemia or sudden cardiac death .Most cases of pulmonary artery hypertension but no atherosclerotic risk factors rarely undergo coronary angiography, and hence, diagnoses are seldom made and proper management is often delayed in these patients.Its natural history is unknown and its treatment has been based more on the severity of the angiographic compression than on the objective demonstration of the myocardial ischemia. In this case report, we describe a patient with severe pulmonary artery hypertension presenting with angina, diagnosed to have extrinsic compression of the left main coronary artery, successfully treated with percutaneous coronary intervention.

Case Report

A 38 –year- woman presented to cardiology outpatient department with history of exertional backache since past 3 months,she also complained of typical postprandial angina but there was no rest angina anytime.She was not a diabetic or hypertensive ,did not have history of hypercholesterolemia or family history of premature coronary artery disease.She reported that she was evaluated 3 years back for progressive shortness of breath and fatigue and was diagnosed to have Congenital heart disease –Ostium secundum atrial septal defect with severe pulmonary artery hypertension with right to left shunt.She had been on phosphodiesterase-5 inhibitors and oral anticoagulation since then and was following up in outpatient regularly. On examination,she had pulse rate of 80 per minute with blood pressure of 100/60 mmHg, had central cyanosis and clubbing.Precordial examination revealed cardiomegaly with wide and fixed second heart sound and accentuated pulmonary component.Electrocardiogram showed sinus rhythm,right axis deviation and right ventricular hypertrophy with systolic strain pattern.Chest X ray showed cardiomegaly, aneurysmally dilated main pulmonary artery (Figure 1). Transthoracic echocardiography showed dilated right sided chambers,aneurysmally dilated main pulmonary artery,large ostium secundum atrial septal defect with right to left shunt (Figure 2).The maximum pulmonary artery systolic pressure recorded was 80mmHg. She underwent MDCT which revealed dilated MPA compressing the ostium of Left main coronary artery causing 90% stenosis. The maximum pulmonary artery diameter was 62 mm. (Figure 3) she underwent Coronary angiogram which confirmed the CT diagnosis. She successfully underwent PTCA to Left Main Coronary artery. (Figure 4) Post procedure was uneventful. Patient was angina free at discharge and remains so on 2 years of follow- up. She continues to be on medications for PAH.

Discussion:

Angina - like chest pain is a common symptom in patients with pulmonary hypertension, occuring in one third of patients during the course of the disease. [7] The causes for this as suggested by McGoon and Cane are as follows 1.Right ventricular ischemia 2.Painful dilatation of the pulmonary artery 3.Extrinsic compression of the left main coronary artery by a dilated pulmonary artery 4.Associated coronary atherosclerosis rarely.

LMCA disease usually occurs secondary to atherosclerotic disease. Other conditions like inflammatory pathologies and extrinsic compression can also lead to LMCA disease. On rare occasions, extrinsic compression of the left main coronary artery is caused by pulmonary artery dilatation. This rare syndrome was first described in 1957 by Corday et al and is characterized by compression of the LMCA between the aorta and an enlarged main PA. (1) It is mostly associated with congenital cardiac defects, mostly an atrial septal defect, ventricular septal defect, or tetralogy of Fallot. [2] The association between an isolated PDA and left coronary artery compression is rare. [3] Other causes are idiopathic pulmonary artery dilatation and primary pulmonary hypertension. These patients have different degrees of pulmonary hypertension, but the determining factor for the compression is the PA dilatation. Risk factors for LMCA compression in patients with pulmonary hypertension include a leftward origin of the left coronary artery, a dilated pulmonary trunk, and an elevated pulmonary trunk to aortic diameter ratio. Both the degree of LM compression and its angle with the left sinus of Valsalva <30% are thought to increase the likelihood of significant myocardial ischemia. A mainpulmonary artery/aorta diameter ratio >2 is considered an additional risk factor. [8]

Compression of the left main coronary artery can be a life –threatening condition. Patients can present with angina, symptoms of left ventricular dysfunction, arrhythmias and/or sudden death. The natural history and its proper treatment remains unclear. [9] Bijl et al reported a patient with extrinsic LMCA compression who suffered a non-Q-wave myocardial infarction and died after 2 days of presentation, other reports however does not demonstrate significant ischemia.[10] In one study by Mitsuda the characteristic narrowing of LMCA was found in 44% of patients with atrial septal defect and pulmonary hypertension. In another study by Kothary et al it is reported in only 4.8%.

Currently, the gold standard for the diagnosis of LMCA compression is coronary angiography with IVUS. Other non-invasive techniques, such as, magnetic resonance imaging and multislice computed tomography(MDCT) may visualize the origin and course of coronaries and allow the detection of the significant coronary artery stenosis. Cardiac 64-slice MDCT provides a noninvasive method for evaluating the degree of LMCA compression, the angulation of the LMCA relative to the left sinus of Valsalva, evaluation of left and right ventricular function, and depiction of pulmonary pathology, making it a valuable investigation in the workup of patients suspected of LMCA compression. Coronary angiography shows a smooth tapering appearance of the LMCA without other evidence of coronary disease suggesting external compression. Both intravascular ultrasound and fractional flow reserve have been validated to guide the treatment of LMCA disease by Pina et al. They recommended that percutaneous or surgical revascularization should be performed if the IVUS minimum lumen area is < 5.9mm and/or abnormal fractional flow reserve is < 0.80.Although coronary angiography is not part of the diagnostic protocol, some authors recommend it to be added to the hemodynamic study of PH patients with symptoms of exertional angina or left ventricular systolic dysfunction.

Compression of the LMCA in patients with pulmonary hypertension is a treatable cause of angina and left ventricular ischemia. Numerous treatment options have been reported, but optimal treatment for this condition remains controversial. These options include percutaneous coronary intervention, coronary artery bypass surgery, and surgical correction of congenital heart disease, pulmonary thromboendarterectomy in PPH and heart lung transplantation.

Treatment of LMCA compression by the pulmonary artery is dependent on the cause and reversibility of the pulmonary trunk dilatation. Fujiwara et al reported the LMCA compression by the dilated pulmonary artery associated with ASD in three patients, one had stenosis of around 75%, two patients had stenosis <50% ,former underwent CABG and latter two patients only ASD patch closure. In the latter two patients, LMCA disease disappeared in postoperative angiograms, suggesting that markedly dilated pulmonary arteries easily compress the LMCA and cause narrowing, which can improve after ASD closure. It is also possible that the pulmonary artery dilatation is not promptly improved because of pulmonary artery remodeling. [7In 2001,Rich et al reported the first 2 cases of patients with PPH and extrinsic compression of the LMCA treated by percutaneous insertion of a intracoronary stent.[1]

Given the high surgical mortality in patients with pulmonary hypertension, LMCA stenting has been favored as the revascularization strategy of choice. Stenting of the LMCA can be carried out with good angiographic results at experienced centers, although too few case histories are available to ascertain its impact on the prognosis of these patients.

Reference

1. Rich S, Dantzker DR, Ayres SM, Bergofsky EH, Brundage BH, Detre KM, Fishman AP, Goldring RM, Groves BM, Koerner SK, et al. Primary pulmonary hypertension: a national prospective study. Ann Intern Med. 1987;107:216–223.
2. Vizza CD, Lynch JP, Ochoa LL, Richardson G, Trulock EP. Right and left ventricular dysfunction in patients with severe pulmonary disease. Chest. 1998;113:576–583.
3. Heikkinen J, Milger K, Alejandre-Lafont E, Woitzik C, Litzlbauer D, Vogt J-F, Klußmann JP, Ghofrani A, Krombach GA, Tiede H. Cardiovocal syndrome (Ortner's syndrome) associated with chronic thromboembolic pulmonary hypertension and giant pulmonary artery aneurysm: case report and review of the literature. Case Rep Med 2012;2012:230736.
4. Andjelkovic K, Kalimanovska-Ostric D, Djukic M, Vukcevic V, Menkovic N, Mehmedbegovic Z, Topalovic M, Tesic M. Two rare conditions in an Eisenmenger patient: left main coronary artery compression and Ortner's syndrome due to pulmonary artery dilatation. Heart Lung 2013;42(5):382–386.
5. Fujiwara K, Naito Y, Iligashine S, et al. Left main coronary trunk compression by dilated main pulmonary artery in atrial septal defect. Thorac Cardiovasc Surg. 1992;104:449–452.
6. Patrat J-F, Jondeau G, Dubourg O, et al. Left main coronary-artery compression during primary pulmonary hypertension. Chest. 1997; 112:842–843.
7. Chest Pain in Association with Pulmonary Hypertension Its Similarity to the Pain of Coronary Disease By

WILLIAM N. VIAR, M.D., AND T. R. HARRISON, M.D.1952. VOL V. NO. 1 8.https://www.ahajournals.org/doi/pdf/10.1161/CIRCULATIONAHA.106.645622

8. Pina Y,ExaireJE,SandovalJ.Left main coronary artery compression syndrome:a combined intravascular ultrasound and pressure wire report.J Invasive Cardiology 2006,18 E102-104.
9. Bijl M, BronzwaerJG, van Rossum AC, VerheugtFW: Angina pectoris due to left main coronary artery compression in Eisenmenger ductus arteriosus. Am Heart J 1993;125:1767-1771.
10. Caldera AE, Cruz-Gonzalez I, BezerraHG, CuryRC, PalaciosIF, CockrillBA, Inglessis-AzuajeI. Endovascular therapy for left main compression syndrome:case report and literature review. Chest.2009;135;1648-1650.

DOI:10.31579/2641-0419/108