A Case of Extrinsic Compression of the Left Main Coronary Artery by Dilated Pulmonary Artery in Eisenmenger 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.

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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 cyanotic 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-old 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

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
main pulmonary 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. [7] In 2001, Rich et al reported the first 2
cases of patients with PPH and extrinsic compression of the LMCA
treated by percutaneous insertion of an 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

7. Chest Pain in Association with Pulmonary Hypertension Its Similarity to the Pain of Coronary Disease By
