CASE REPORT

LEFT MAIN CORONARY ARTERY DISEASE IN PATIENTS WITH ATRIAL SEPTAL DEFECTS (ASD)

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CASE # 1

38 year old woman was admitted in National Institute of Cardiovascular Diseases (NICVD), with complaints of typical chest pain and progressive exertion dyspnoe, for last 5 years, currently she was in NYHA 3 shortness of breath. Physical examination revealed RV heave, right ventricular s3 and fixed splitting of second heart sound in all phases of respiration and grade 111/V1 systolic murmur at right second intercostal space. ECG showed sinus rhythm with right axis deviation and signs of right ventricular hypertrophy and negative T waves in anterior and inferior leads. Chest x-ray showed cardiomegaly with dilated pulmonary arteries. ECHO showed large Secundum Atrial Septal defect (ASD), moderate TR and estimated pulmonary pressure of 90 mmHg. The patient underwent cardiac catheterization which revealed oxygen step up at the level of right atrium, pulmonary pressures was 80/25, mean PA pressures was 45 mmHg, QP/QS was 2.4. Although she had no coronary risk factors coronary angiography was planned due to her complaints of shortness of breath which was out of proportion to her underlying disease and her complaints of chest pain, angiography showed severe ostial left main disease (fig 1 & 2) while rest of the coronary arteries were free of disease. She was referred for ASD closure along with graft to LAD.

CASE # 2

55 years old female, a known case of ASD underwent coronary angiogram before surgical closure of ASD with PAH, due to NYHA 3 shortness of breath. Her...
Coronary angiogram showed severe ostial left main disease (fig 3 & 4), while rest of her coronaries were normal. She was referred for ASD closure and graft to LAD.

**CASE # 3**

50 years old male, current smoker was evaluated in OPD for progressive shortness of breath since last 6 months, his physical examination was consistent with typical findings of ASD, echo confirms Secundum ASD with pulmonary hypertension, and QP/QS of 2.7. Coronary angiogram revealed severe distal Left main disease with severe tubular mid LAD disease and severe discrete OM2 disease (fig 5 & 6). He was referred for combined CABG and ASD closure.

**COMMENTS**

Coronary artery stenosis is usually caused by atherosclerotic disease. However, on rare occasions, extrinsic compression of the left main coronary artery (LMCA) is caused by pulmonary artery dilatation. The extrinsic compression of left main coronary artery(LMCA) secondary to pulmonary artery trunk dilatation is a relatively newly described syndrome that has been associated with severe pulmonary hypertension. It is primarily related to congenital heart disease or idiopathic pulmonary hypertension but other causes of pulmonary hypertension can be the triggering factor for this syndrome. Prolonged left-to-right shunt in patients with ASD promotes right heart dilatation and pulmonary vascular changes. Extrinsic compression of the LMCA by an enlarged pulmonary trunk in patients with long-standing pulmonary hypertension has been previously reported. The cause of ostial LMCA stenosis needs to be evaluated in a patient with long standing severe pulmonary hypertension for possible extrinsic compression of the LMCA secondary to dilated pulmonary artery. In 1957, Corday et al first described the compression of the LMCA by a dilated pulmonary artery as the cause of coronary insufficiency in patients with pulmonary hypertension. These patients have different degrees of pulmonary arterial hypertension, but the determining factor for the compression of the LMCA is the PA dilatation. This condition may have a potential role.
in the genesis of angina, left ventricular dysfunction, arrhythmias and/or sudden death in patients with pulmonary arterial hypertension. The natural history of this disease and its proper treatment, however, remain unclear.7 Bijl et al reported a patient with extrinsic LMCA compression suffered a non-Q wave myocardial infarction and died after 2 days of presentation, other reports however does not demonstrate significant ischemia.8 The optimal management of LMCA compression caused by a dilated pulmonary artery secondary to ASD is yet to be clarified. Some authors have proposed aggressive treatment such as coronary artery bypass surgery or stent implantation.1,3-15 However, the benefit of coronary revascularization remains controversial because simple surgical closure of ASD may relieve the LMCA compression by lowering the pulmonary artery pressure and resultant decrease in pulmonary artery diameter. Fujiiwara and colleagues3 reported that the LMCA compression by the dilated pulmonary artery associated with ASD in three patients, one had LMCA stenosis of around 75% while rest of two patients had LMCA disease < 50%. The former underwent CABG with ASD closure while latter two patients referred only for patch closure of ASD. In the latter two patients LMCA disease disappeared in postoperative coronary angiograms. These results suggest that markedly dilated pulmonary arteries easily compress the LMCA and cause narrowing, which can improve after ASD closure. On the other hand, it is possible that the pulmonary artery dilatation is not promptly improved after ASD closure because of the pulmonary artery vascular remodeling caused by chronic pulmonary artery hypertension, and as a consequence, the degree of LMCA compression may not change. As extrinsic compression of LMCA may cause angina and lead to malignant arrhythmia or sudden death, a precise assessment for LMCA narrowing is very important.16 Both intravascular ultrasound and fractional flow reserve have been validated to guide the treatment of left main coronary artery disease by Pina et al. They recommended that percutaneous or surgical revascularization should be performed if the intravascular ultrasound minimum lumen area is < 5.9 mm and/or abnormal fractional flow reserve is < 0.80.7 Given the high surgical mortality in patients with pulmonary hypertension, LMCA stenting has been favored as the revascularization strategy of choice. In 2001, Rich et al reported successful LMCA stenting in two patients with primary pulmonary hypertension and LMCA compression syndrome6 Since then, several other authors10-15 have reported successful angiographic and short-term clinical outcomes with this technique. Of note, all of the reported cases have involved compression of the ostium or proximal LMCA, sparing the left main bifurcation, and single stent placement was required. In the present cases, although the patient # 1 did have typical angina with negative troponin, with her electrocardiography revealed negative T waves in precordial and inferior leads and isolated LMCA ostial disease on coronary angiogram, we decided to perform the bypass surgery for LMCA compression as a safety and due to financial constraint, because it was not certain whether the compression would promptly disappear by ASD closure in a short period of time. Similarly patient #2 also underwent CABG with ASD closure for the same reasons. However patient # 3 had atherosclerotic narrowing of LMCA and significant lesions in mid LAD so referred for combined ASD closure and CABG. Advances in both noninvasive and invasive imaging have facilitated the recognition and management of patients with this condition. Cardiac 64-slice, multi-detector CT scanning provides a comprehensive method for evaluating the degree of LMCA compression, the angulations of the LMCA relative to the left sinus of Valsalva, the evaluation of left and right ventricular function, and pulmonary pathology both before surgery and monitoring post surgical follow up.17

In conclusion, there are several teaching/learning prospectuses from above cases.

A. LM disease in patients with pulmonary hypertension should be carefully evaluated for either true atherosclerotic obstruction or external compression from dilated pulmonary arteries.

B. Due to high operative mortality of patients with pulmonary hypertension, hybrid procedures involving stenting of Left main coronary artery and surgical closure of ASD should be considered.

C. Although no definite guidelines for treatment of isolated LMCA with pulmonary dilatation due to PAH are available but advanced pre-treatment modalities like CT scan, IVUS & FFR are now available in our set up, so from now on one should
used all available modalities before initiating treatment to minimize intra-operative risk and mortality and in the follow up for such patients.

REFERENCES


