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Case Report

Internal Medicine Section

Early Post-Operative Coronary Thrombosis Following Repair of a Proximal Coronary Artery Fistula

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ABSTRACT

Patients with aneurysmal coronary artery fistulas are often a treatment challenge. We hereby, report a case of aneurysmal left main coronary artery to coronary sinus fistula repair, complicated by an early post-operative thrombosis of the left main coronary artery, necessitating an orthotropic heart transplant. Routine use of peri-procedural and long-term anti-coagulation is usually not a standard recommendation in these cases; however, early institution of the same may prevent flow stasis, thrombus formation and unfavourable outcomes pre- or post-operatively.

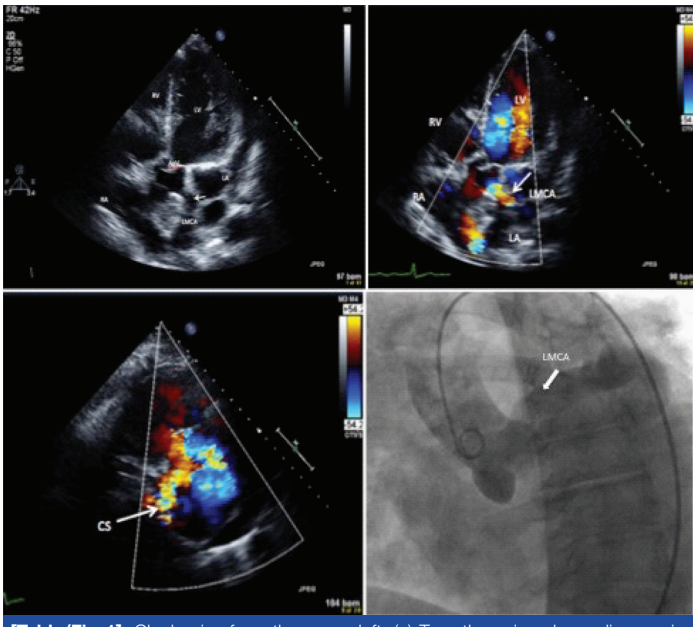
Keywords: Coronary artery aneurysm, Heart transplant, Thrombus formation

CASE REPORT

A 59-year-old female with a known congenital Left Main Coronary Artery (LMCA) to Coronary Sinus (CS) fistula, presented with worsening dyspnoea. The patient refused to consider any intervention previously. Physical examination revealed a continuous murmur over the precordium and evidence of volume overload.

Further evaluation with a Transthoracic Echocardiogram (TTE) revealed dilated cardiac chambers, a Left Ventricular Ejection Fraction (LVEF) of 35% which was 60% two years ago, dilated CS and proximal LMCA, with evidence of left to right shunting between them [Table/Fig-1a-c]. Subsequent coronary angiogram showed a dilated LMCA [Table/Fig-1d]. A right heart catheterisation revealed a step-up in oxygen saturation in the right atrium (Qp/Qs 1.8).

The patient underwent a surgical ligation of the fistula at its origin close to the LMCA. She was stable immediately post-operatively; however, on post-operative day 1, she developed cardiogenic shock (EF 15%); an urgent coronary angiogram revealed complete thrombosis of the LMCA, necessitating emergent coronary artery bypass grafting. Subsequently, she required support with Extra-Corporeal Membrane Oxygenation (ECMO). On post-operative day 7, she failed ECMO clamping and underwent left ventricular assist device implantation. In the following months her clinical course was complicated by repeated heart failure re-hospitalization. She eventually underwent successful orthotopic heart transplantation.



[Table/Fig-1]: Clock wise from the upper left: (a) Transthoracic echocardiogram in the apical 5-chamber view showing dilated chambers and severely dilated left main coronary artery ostium (arrow); (b) Color flow doppler revealing increased flow across the ostium (arrow); (c) Coronary sinus (arrow); (d) Aortogram of dilated proximal left coronary artery (arrow).
RA-right atrium, LA-left atrium, LV-left ventricle, RV-right ventricle, LMCA-left main coronary artery, CS-coronary sinus, AoV-aortic valve

DISCUSSION

Coronary Artery Fistulas (CAF) are abnormal connections between coronary arteries and a cardiac chamber or major vessel [1-3]. They are the second most common congenital coronary anomaly, with an incidence of 0.2% to 0.6% in patients undergoing coronary angiograms [4]. Guidelines for management of adults with congenital heart disease [4] recommend closure of all large CAFs or symptomatic CAFs irrespective of their size [5]. Large fistulas (>three times the normal proximal coronary artery diameter) or shunts (Qp/Qs >1.5) are haemodynamically significant and often symptomatic [1,4].

Coronary steal often leads to myocardial ischaemia, which in turn causes compensatory dilation of the native coronary artery with possible aneurysmal formation [1].

Surgical closure is preferred for CAFs with the following characteristics: i) locations out of reach for percutaneous intervention; ii) multiple fistulas; iii) extreme native vessel tortuosity; (iv) aneurysmal dilation of fistula or proximal coronary artery; iv) presence of normal coronary branches near the fistula's drainage sites [1].

Transcatheter closure alternatively is considered the procedure of choice, given its shorter recovery time for patients; however, it is limited to smaller and anatomically simpler fistulas [1].

Several studies have reported early and late symptomatic coronary thrombosis after surgical and percutaneous repair of CAF in adults [Table/Fig-2] [5-10]. Distal CAFs [8-10] often are at greater risk of flow stasis and thrombosis following closure, due to greater proximal coronary artery dilation [1]. Whereas, proximally located fistulas, such as in our case, may have formed a blind pouch between the ligated portion of the fistula and the native coronary artery following fistula ligation, leading to thrombus propagation [5-7].

Fistulas found draining into the coronary sinus tend to have poorer outcomes compared to those draining into the right ventricle, pulmonary arteries or left ventricle [1,11]. It is believed that, following surgical repair there is risk of extension of a thrombus

Reported Case (reference)	Age at Diagnosis (years), Sex	Origin, Termination of CAF	Location and Size of CAF	Associated Coronary Artery Dilation	Treatment Pathway	Timing of Thrombosis	Coronary Anatomy	Outcome	Mortality
Mesko ZG et al., [5]	4, Female	LMCA, RA	Proximal, Large	-	Surgical	Late (11 years)	LAD and D-1 bifurcation with thrombus	Normal LV function and Coronary Flow	Nil
Toyoda Y et al., [6]	10, Female	RCA, RA	Proximal, Large	-	Surgical	Late (day 9)	Thrombus in dilated proximal segment	Normal LV function	Nil
Goldberg SL et al., [7]	37, Male	LCX, CS and RCA, CS	Proximal, Large and Distal, Small	LMCA, LCX	Surgical	Late (1.5 years)	Occluded proximal LCX	LCX thrombosis with stent placement (1.5 years), OM occlusion requiring CABG (2 years)	Nil
Ascoop AK et al., [8]	69, Female	RCA/RCC, RA	Distal, Large	Proximal RCA	Percutaneous closure, Amplatzer VSD occluder	Early (few hours)	Thrombus in blind pouch of fistula/ proximal RCA	Hemodynamic Collapse, CABG	Nil
Kharouf R et al., [9]	48, Female	LCX, CS	Distal, Large	LMCA, LCX	P, Coil Embolisation	Late (4 days)	Thrombsis of complete LCX	LV posterio-inferior wall hypokinesis, Intermittent Atrial fibrillation, Pulmonary Embloism	Nil
Hamada M et al., [10]	26, Male	LAD, RV	Distal, Large	LAD	Surgical	Late (9 months)	Thrombsis of LAD	-	Nil
Current Case	59, Female	LMCA, CS	Proximal, Large	RCA, LCA	Surgical	Early (1 day)	Thrombsis of complete LCA	Cardiogenic Shock, LVAD, OHT	Nil

[Table/Fig-2]: Summary of studies reporting adverse coronary events after proximal or distal coronary fistula closure.
*LMCA (left main coronary artery), RA (right atrium), RCA (right coronary artery), RCC (right coronary cusp), S (surgical closure), P (percutaneous closure), LV (Left Ventricle), LCX (left circumflex artery), D-1 (First Diagonal artery from left anterior descending artery), LCA (Left Coronary Artery), LVAD (Left Ventricular Assist Device), OHT (Orthotopic Heart Transplant), OM (obtuse marginal), CABG (coronary artery bypass grafting).

from the dilated coronary sinus into the major epicardial artery [11].

Further, patients who have delayed coronary thrombosis may have better outcomes, compared to those with early thrombosis, as their opposing coronary system can slowly adapt by increasing collateralization [12].

In our case, the patient had a congenital proximal LMCA fistula that led to significant coronary dilation with risk of stasis of blood flow when the shunt was closed.

CONCLUSION

Consensus on preventing such thrombi is lacking. The decision to start anti-platelet/anti-coagulation therapy in patients following CAF repairs may be determined by the size of the fistula, presence of residual aneurysms and by balancing the patient's risk of bleeding with thrombosis.

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