1. Abstract

Coronary cameral fistula is a sizeable communication between a coronary artery, bypassing the myocardial capillary bed and entering a chamber of heart. It constitutes 0.2%-0.4% of congenital cardiac anomaly. Sixty percent of these fistula arise from the right coronary artery and terminate mostly (90%) in the right side of the heart. The most frequent sites of termination in the descending order are, the right ventricle, right atrium, coronary sinus, and the pulmonary vasculature.

2. Introduction

We present a case of a young female who was symptomatic and was evaluated for the same. The fact that she had a coronary cameral fistula from the left main coronary artery terminating into the Superior vena cava and right atrium junction and was operated successfully on empty beating heart without arresting heart, avoiding cardioplegia and cardiotomy made the case unusual one.

3. Case Presentation

28yrs female who was asymptomatic previously presented to us with atypical chest pain, palpitation and breathlessness on exertion of 6mth duration. Her cardiovascular examination suggested loud second heart sound & continuous murmur in the precordium. Electrocardiogram & blood investigations were within normal limits. Xray was suggestive of cardiomegaly. Echo showed good LV function. Coronary angiogram showed an aneurysmally dilated LMCA with a fistulous tract terminating in the superior vena cava and right atrium junction. Patient was referred for surgery as it was unsuitable for transcatheter closure. Patient was intubated and trans esophageal echocardiogram (TEE) probe was put to confirm the findings. Midline sternotomy was done and findings were noted. Right atrium was dilated, pink in color and there was obvious thrill was palpable at superior vena cava and right atrium junction. Patient was taken into cardiopulmonary bypass using usual standard technique. Fistula was dissected, traced and doubly (Figure 1) ligated and transfixed at its origin in empty beating heart avoiding cross clamping and cardioplegia. The thrill and murmur disappeared immediately after ligation which was confirmed by TEE. Pt. was extubated within few hrs of surgery. Precordial continuous murmur completely disappeared Post op. echo was normal. Patient was discharged on 5th post-op day. After two years of surgery she is completely asymptomatic and following up with us.

4. Discussion

Coronary fistulae account for 0.2 to 0.4% of the congenital cardiac abnormalities, and about 50% of the paediatric coronary vasculature abnormalities. It may appear as a persistence of sinusoidal connection between the lumen of the primitive tubular heart that supplies the myocardial blood flow in the early embryonic period [1,2]. It also occurs in 45-50% of patients.
with pulmonary atresia, who have intact ventricular septum. Their morphogenesis is thought to begin early in the development with embryological myocardial lacunae when myocardial blood supply is derived from its own lumen. Subsequently the coronary arteries communicate with these intramyocardial trabecular spaces. These sinusoids may involute forming the microcirculation. If the pulmonary valve fuses before the sinusoids contract, RV pressure increases from isovolumic contraction and flow into the sinus is maintained. The pathophysiological mechanism of coronary fistula is myocardial stealing or reduction in myocardial blood flow distal to the site of connection [3]. The coronary vessel tries to compensate by progressive enlargement of the ostia and the feeding artery. This explains the aneurysmal malformation of the LMCA and LCx arteries in the present case. With time, these arteries leading to the fistula may progress to frank aneurysm, intimal ulceration, intimal rupture, atherosclerotic deposition, calcification, and mural thrombosis. It may rarely rupture. It has also been proposed that a congenital coronary artery fistula proximal to a segment of acquired atherosclerotic stenosis aggravates the distal perfusion deficit by acting as a low resistance alternative to the zone of coronary artery stenosis is [4]. The anaesthetic management of these patients is usually centred around the prevention of coronary steal and perioperative myocardial ischaemia, which can occur with an increased left to right shunt. Hence Cardiopulmonary bypass (CPB) was established using standard techniques. And fistula was dissected, doubly ligated and transfixed on empty beating heart avoiding cardioplegia and opening of any chamber of heart. During ligation flow was reduced and assistant was retracting and pressing the aorta to assure complete obliteration of fistulous tract.

Interventional catheterization is yet another therapeutic option available for treatment. The technique uses coils or other devices and can be performed on an outpatient basis. Fistulae unsuitable for trans-catheter approach include those with multiple connections, circuitous routes and acute angulations that make catheter positioning difficult or impossible [2]. Coil embolisation was not considered in the present patient, as it was big and aneurysmal. Spontaneous closure occurs in 23% of small fistulae, primarily those arising from left coronary system.

5. Conclusion

Coronary cameral fistula from LMCA to SVC-RA junction is very rare and unusual. Surgery was successfully done on pump beating heart without arresting heart avoiding cardioplegia and cardiotomy makes it interesting one.

Reference


