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An Intramural Left Main Coronary Artery with a Left Sinus of Valsalva Aneurysm: A Unique Combination of Congenital Anomalies

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ABSTRACT

The congenital anomaly of an intramural left main coronary artery arising in the anatomically correct aortic sinus is very infrequent. Aneurysms involving the sinus of Valsalva rarely arise from the left aortic sinus. We present the clinical features and surgical correction of this rare anomaly along with a short discussion of these congenital malformations.

INTRODUCTION

The congenital anomaly of the intramural left main coronary artery (LMCA) arising in the anatomically correct aortic sinus is very infrequent [Angelini 2010]. Aneurysms involving the sinus of Valsalva rarely arise from the left aortic sinus. We present the clinical features and surgical correction of this rare anomaly along with a short discussion of these congenital malformations.

CASE REPORT

A 57-year-old man presented to the emergency department with unstable angina. Given his significant risk factors (hypertension, diabetes, smoking), we suspected coronary artery disease as the etiology of his chest pain. The coronary angiogram was normal, however, and the transthoracic echocardiogram revealed an unruptured small left sinus of Valsalva aneurysm (SOVA) without compression of the LMCA. The aortic valve was normal, and the left ventricular ejection fraction was preserved. A computed tomogram of the chest was done to determine the anatomic relationship of the SOVA to surrounding structures (Figure 1). This evaluation revealed that both coronary ostia arose from the midpoint of their anatomically correct sinuses with respect to the commissures, but they were displaced cephalad, arising near the sinotubular junction. The LMCA had an obliquely directed intramural course. We chose to repair the SOVA and inspect the LMCA ostium, because we suspected ostial narrowing as the cause of his angina.

After opening the aorta, we located the unruptured left SOVA (1.3x1.8 cm), which had a distinct neck. It was closed primarily with interrupted Prolene sutures. Both coronary ostia were present in the middle of the appropriate sinuses with respect to the commissure, but they were displaced cephalad closer to the sinotubular junction. The orifice of the LMCA was severely narrowed and reduced to a slit-like opening. Using an intracoronary dilator as a guide, we carefully excised the common wall to create an obstructed opening into the distal LMCA. The residual intimal layers were approximated with 6-0 polypropylene sutures (Figure 2). We anastomosed the distal left anterior descending artery to the harvested left internal mammary artery to further protect the left coronary system.

DISCUSSION

SOVA is a rare cardiac anomaly with an estimated incidence of 0.09% to 0.15% in the general population, with left SOVA being extremely rare [Schullerer 2011]. Myocardial ischemia in these patients is generally due to local coronary artery compression or high-output congestive heart failure secondary to intracardiac rupture of the SOVA. Our patient had a small, intact SOVA; hence, the myocardial ischemia could not be attributed to this anomaly. We thus chose to perform a computed tomography evaluation to further investigate the cause of the ischemia. This investigation was the only one that revealed the actual diagnosis. Computed tomography been established as the modality of choice in the investigation of coronary anomalies.

An LMCA originating from the normal left sinus with a proximal intramural course was first reported in 2010 [Angelini 2010]. Vasospasm is the most common cause of myocardial ischemia and may even lead to sudden cardiac death. This mechanism of ischemia is unlikely in our patient, however, because the proximal LMCA lacks the tunica media [Angelini 2010]. Rather, the presence of a membrane between the intramural LMCA and the aorta produces a compromised luminal diameter and a reduced blood flow in the LMCA. The stiff membrane hampers the diastolic relaxation of the coronary ostium, further compromising the coronary blood flow.

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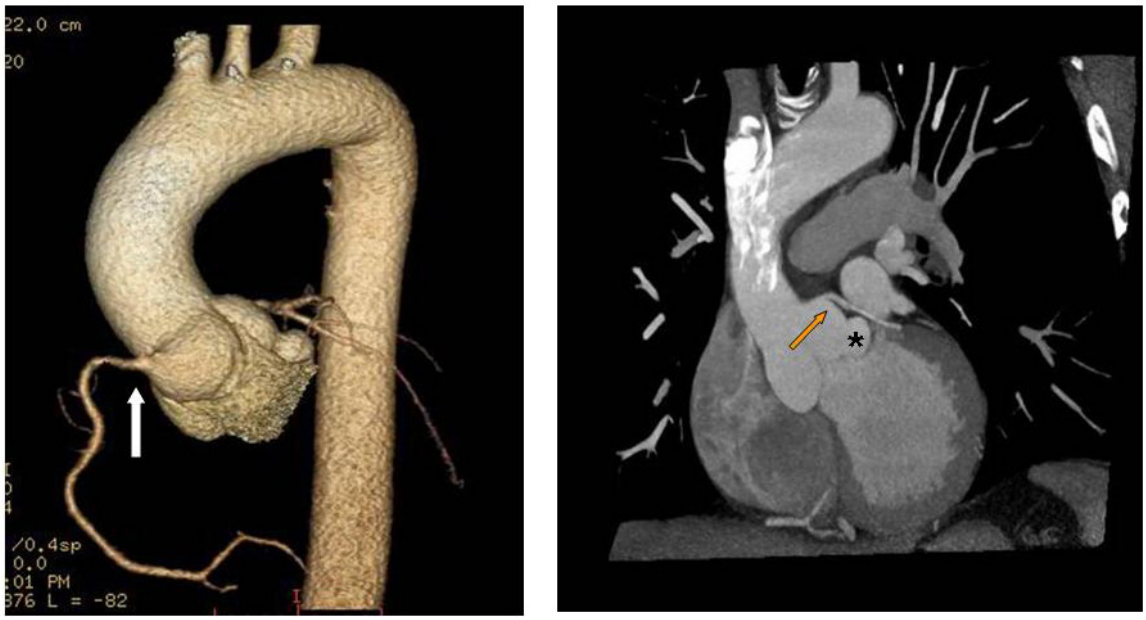

Figure 1. Left, A reconstructed computed tomogram depicting the anomalous high origin of the left main coronary artery (arrow) from the left coronary sinus. Right, Computed tomographic angiography image showing the high origin of the left main coronary artery being pushed upward (arrow) by the left sinus of Valsalva aneurysm (*).

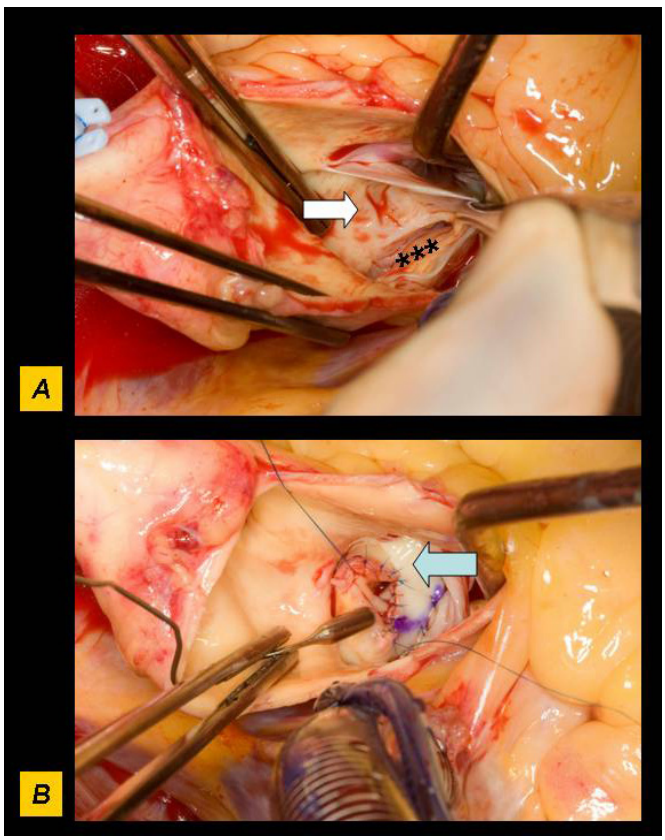

Figure 2. A, Intraoperative photograph showing the small left sinus of Valsalva aneurysm (***), along with the narrowed ostium of the left main coronary artery (arrow). B, The ostium of the left main coronary artery is now wide open at the conclusion of the surgical procedure (arrow).

Sudden cardiac death or myocardial ischemia can occur if the membrane narrows the ostium by >50% [Frescura 1998].

Our case presents a combination of 2 rare congenital anomalies of the sinotubular area. The aneurysmal expansion of the left coronary sinus would increase the distance between the aortic annulus and the sinotubular junction, thereby shifting the ostium of an LMCA cephalad and further increasing the length of the intramural segment. It is debated whether the catenoidal configuration of the aortic sinuses determines which 2 would develop coronary buds [Hutchins 1988]. Given that SOVA is basically an anomaly of the medial layer of the aortic sinuses, we speculate that the presence of a SOVA could have led to the development of the intramural LMCA. That may also explain why the right coronary artery was not intramural, despite having an equally high origin in its respective coronary sinus.

Surgical management of coronary anomalies has to be tailored to the anatomic abnormalities encountered. The coronary orientation was normal in our patient, and therefore coronary transfer was not required. Excision of the ostial membrane created an adequately sized orifice, and we were confident that adequate left main coronary flow would resume.

CONCLUSION

The presence of an intramural LMCA and a left SOVA in the same patient is unique, but they may have a common embryologic origin. The surgical management of coronary anomalies should be directed toward simple techniques that can achieve an unobstructed coronary blood flow.

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