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Short Communication

Surgical Correction of Malignant Right Coronary Anomalies Stranding an Aortic Commissure with and without Unroofing

Khaled Abdelhady, Samarth Durgam M, Chawki Elzein, Michel N. Ilbawi, David Rhoiney DO, Malek G Massad*

Department of Surgery and Cardiothoracic Surgery, University of Illinois at Chicago, USA

***Corresponding author**

Malek G. Massad , Department of Surgery, University of Illinois at Chicago, 840 S Wood St, CSB Suite 417(MC 958), Chicago, IL 60612, Division of Cardiothoracic Surgery, University of Illinois, USA, Tel: 1312-996-4942, Email: mmassad@uic.edu

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Abstract

The technique for successful surgical correction of an anomalous origin of the right coronary artery from the opposite aortic cusp with a malignant course between the aorta and pulmonary artery is illustrated in a symptomatic 62 year old female patient. The malignant intramural course of the right coronary artery traversed the tip of the commissure between the anterior and posterior leaflets and its repair entailed unroofing of the intramural segment from inside the aortic intima. This technique required resuspension of the overlying commissure to maintain optimal aortic valve leaflet cooptation and prevent aortic insufficiency. Modifications of this technique have been utilized by us whenever the intramural segment traversed behind the commissure. In these cases, partial or subtotal unroofing of the intramural segment was performed to preserve the integrity of the intima behind the overlying commissure. More recently we have performed the surgical correction by probing the intramural segment within the aortic wall through the anomalous origin to its most anterior location then performing a wide anterior unroofing in the aortic intima and marsupial zing the aortic and coronary intima to avoid dissection or intimal flap development. We favor utilizing these techniques of anatomic correction of the anomalous coronary to other techniques involving coronary artery bypass grafting of the anomalous coronary especially in adult patients as unroofing provides more lasting results.

INTRODUCTION

Malignant right coronary artery anomalies remain a challenging problem to manage both in the pediatric and adult age groups. Aberrant origin of the right coronary artery from the left coronary sinus and coursing intramurally between the ascending aorta and main pulmonary artery to take its normal anatomic course towards the right atrioventricular groove is one of the anomalies that have been recognized. The incidence is 0.05% to 1.2% according to autopsy, angiographic or prospective echocardiography findings [1]. Myocardial ischemia, syncope, and sudden cardiac death commonly in athletes have been associated with this anomaly [2]. The surgical treatment options for this type of right coronary anomaly include an anatomic correction of the anomaly in all age groups or, as another option in the adolescent and adult patients, performing a coronary artery bypass using an internal mammary artery or a saphenous vein graft [4]. The latter has its limitation since the obstruction is often dynamic and the coronary bypass can lead to conduit graft occlusion due to competitive flow and may have limited durability due to thrombosis, neointimal hyperplasia or arteriosclerosis [4].

The standard approach for repair entails unroofing of the intramural coronary segment within the aortic wall thereby transposing the posteriorly located anomalous origin to an anterior location where the neo-ostium and proximal segment have a perpendicular trajectory from the aorta rather than an intramural course. This approach for correction can prove difficult when the anomalous course within the wall of the aorta traverses and strands an aortic commissure overlying the intramural segment, which is often the case. As a result, repairs that include unroofing can occasionally lead to a floppy aortic valve due to commissural mal-alignment and aortic insufficiency [4].

In this communication, we report on the surgical technique and its modifications that were used to complete the correction and highlight some of the challenges in the repair that can overcome to avoid the development of aortic insufficiency.

Surgical technique

The standard technique for anatomic correction was utilized in this index case and involved re-suspension of the overlying commissure by attaching it back to the aortic wall using pledgeted polypropylene (Prolene) sutures to maintain the structural integrity of the aortic valve. A 62 year old woman presented to our center with angina pectoris and had a positive nuclear stress test indicating ischemia in the territory of the right coronary artery. A coronary angiogram showed an aberrant origin and course of her right coronary artery with the right coronary ostium originating from a separate ostium within the left coronary cusp (Figures 1,2). The right coronary artery had a malignant intramural course traversing the commissure between the left and right aortic valve leaflets and within the aortic wall between the ascending aorta and main pulmonary artery. Surgical correction was performed with standard cardiopulmonary bypass and left ventricular venting. After cross clamping the aorta, we exposed the aortic root through a hockey-stick incision, an inch above the commissures. The left and right coronary ostia were identified within the left coronary cusp. A fine metallic probe with an olive tip was passed through the right coronary ostium to delineate the trajectory within the aortic wall. In this case, the intramural segment traversed the tip rather than the actual body of the commissure separating the anterior and posterior aortic valve leaflets. The intimal wall of the intramural segment is then slit open from the anomalous ostium all the way behind the commissural tip to allow unroofing of the intramural tract with a number 11 blades and a fine Potts scissor. After unroofing the trajectory throughout the length of the intramural segment, redundant parts of the intima forming the roof of the intramural segment were excised and the two walls of the trajectory were marsupial zed by re-approximating them with interrupted sutures of 6-0 Prolene.

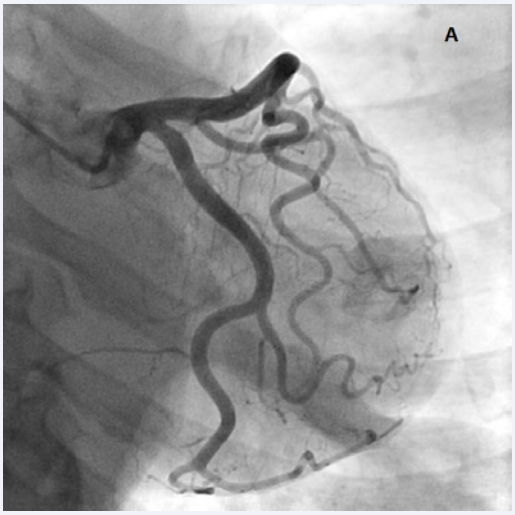

Figure 1 Angiogram showing the normal course of the left coronary artery.

The new right coronary ostium created is now transposed to the right thereby avoiding aortopulmonary compression of the now unroofed intramural right coronary segment (Figure 3). The neo-ostial intima is, likewise, fixed with interrupted 6-0 Prolene sutures to avoid dissection or extravasation. Once this has been accomplished, the commissure between the left and right coronary ostia is resuspended back to its original location on the aortic wall to preserve the length of the Sino-tubular junction and avoid floppiness of the aortic leaflets. This was accomplished using a 5-0 Prolene suture with Teflon pled gets

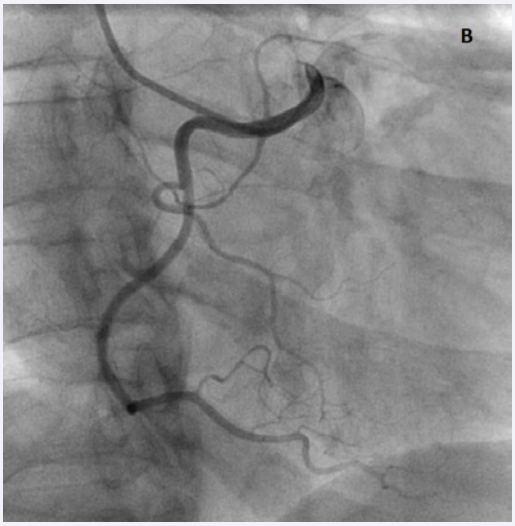

Figure 2 Angiogram showing the anomalous right coronary artery from the left coronary artery cusp.

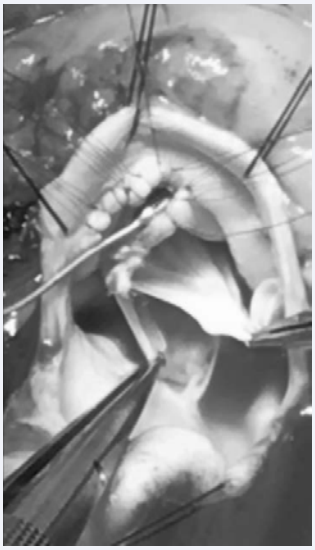

Figure 3 An intraoperative view of the aortic root showing the corrected origin and course of the anomalous right coronary segment. The intramural coronary artery is unroofed and marsupialized and the new right coronary ostium is transposed back into its original location. The commissure between the anterior and posterior aortic valve leaflets is resuspended by suturing it back to the aortic wall to preserve the sinotubular junction and maintain optimal aortic leaflet coaptation.

on the outside and inside of the aortic wall through the tip of the commissure. The integrity of the repair is evaluated with saline irrigation of the aortic valve confirming optimal cooptation of the aortic leaflets with no residual leak. The aortic incision for exposure is then closed in two layers of 4-0 Prolene sutures. The patient is warmed back to normothermia and separated from cardiopulmonary bypass. Intra-operative trans-esophageal echocardiography showed normal left ventricular wall motion with no residual aortic insufficiency and an electrocardiogram performed prior to hospital discharge on the fifth postoperative day showed normal sinus rhythm with no ST segment depression or elevation. The patient was asymptomatic and doing well on follow up six months after the operation.

COMMENTS

The anomalous origin of the right coronary artery from the left aortic sinus coursing between the great vessels has been recognized as a potential risk for chest pain, syncope and sudden death [5]. On the contrary, the patient can also be asymptomatic when either left or right anomalous coronary artery coursing between the great vessels, and are diagnosed only at autopsy [1]. Although there is no consensus regarding the management, a surgical correction is being considered in view of large evidence that these patients are prone to experience myocardial ischemia that could potentially lead to sudden death [4,6-8]. The pathophysiologic mechanism of ischemia in these patients was originally postulated to be a result of exercise induced compression of the anomalous coronary artery by the pulmonary artery on one side and the aorta on the other [3]. This mechanism has been challenged lately as unlikely to be the cause because of the low pressure in the pulmonary artery compared to the coronary pressure. Instead, it is now thought that myocardial ischemia occurs due to an abnormal angulation of the origin of the anomalous coronary that is exacerbated by aortic distention such as during exercise with acute stretching of the intramural segment thereby leading to luminal narrowing [8].

Re-implantation of the anomalous coronary artery into the correct sinus of the aorta, unroofing of the intramural segment of the coronary artery, main pulmonary artery translocation, and coronary artery bypass grafting are the various techniques currently used in the management of coronary artery anomalies [6,9,10]. Coronary artery translocation and direct aortic re-implantation is one of the preferred surgical approaches and can be performed safely in most cases [9,10]. However, single coronary artery or insufficient lengths after mobilization to re-implant are the limitations to carry out direct re-implantation [9]. In such scenario, pulmonary artery translocation can be the alternative option which was first described by Rodefeld et al or a coronary artery bypass grafting. In 2002, we reported our experience with 8 patients who had right or left coronary artery anomalies that was repaired of which 6 patients had their right coronary artery originate from the left coronary sinus with a malignant course between the aorta and pulmonary artery [11]. The surgical correction included unroofing of the intramural coronary segment in all patients. Since then, our experience has grown to 27 patients with coronary anomalies that were all corrected successfully. Their age ranged from 2 months to 62 years. All patients are asymptomatic and doing well at a mean follow up of 15 years with no morbidity and mortality attributed to the repair.

The unroofing procedure since reported by Mustafa et al, is currently the approach of choice showing satisfactory results [12]. However, aortic insufficiency remains as one of the expected complications of this repair due to detachment of the inter-coronary commissure [13]. In order to avoid the potential aortic valve incompetence, a modified technique with creation of neo-ostium without or partial unroofing has been implemented [13,14]. At the beginning of our experience we had one patient who developed intraoperative mild to moderate aortic insufficiency, in this case we re-suspended the commissure after unroofing and this required no further surgical intervention. We attributed that to possible shortening of the sinotubular segment during commissural resuspension or possible detachment of the resuspended commissure. We have since performed surgical corrections using a modification of the above technique whenever we were faced with an intramural segment traversing the middle rather than the tip of the overlying commissure. In these instances, we performed either subtotal unroofing or tracheotomy preserving only the sub-commissural intima or in some cases performed partial unroofing of the part of the intramural segment anterior to the commissure. In other instances, we completely eliminated the need for unroofing of the intramural segment of the coronary artery. Instead, we were able to probe the intramural tract through the anomalous coronary ostium to its most anterior aortic wall location and then transpose the ostium to its supposed most anterior location by performing a wide incision in the intima of the aortic wall at the tip of the probe. The split aortic intima and the coronary intima at the new origin of the right coronary artery are then re-approximated to create the neo-ostium. There were no other surgical complications that were attributed to the procedure and no operative or postoperative deaths that occurred among patients who had these modified techniques.

As we have accumulated a growing experience with this anatomic correction among pediatric and adult patients with malignant coronary anomalies, we propose utilizing this technique or its modifications instead of coronary graft bypass in the adult patient population. However, coronary artery bypass is a valid option in patients with co-existing atherosclerotic coronary artery disease [13]. It is clear that an anatomic correction of the right coronary artery origin anomaly would provide a more lasting result than coronary artery bypass with arterial or venous conduits due to the several limitations that were previously described.

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