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A Rare Case Report on Coronary Artery Anomaly: from Embryology to Emergency

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Abstract: ***Anomalous coronary artery origin is a rare congenital cardiac abnormality that can have significant clinical implications, particularly when associated with a "malignant" course. Among these anomalies, the anomalous origin of the Left Coronary Artery is particularly critical due to its vital role in myocardial perfusion. This condition is especially concerning when the Left Coronary Artery takes an interarterial course between the aorta and pulmonary artery—a trajectory termed "malignant" due to its association with sudden cardiac death (SCD), particularly in young athletes. Hereby, we present a case of a 73 year old male who presented with breathlessness on exertion since 1 month with a positive TMT. CT Coronary Angiography study revealed Anomalous origin of Left Main Coronary Artery (LMCA) from Right Coronary Artery (RCA) with a malignant course between the aorta and pulmonary artery followed by an intramural course. The study also showed Double Vessel Disease affecting the Left Circumflex Artery (LCx) and Right Coronary Artery (RCA).***

Keywords: congenital cardiac abnormality, malignant course, sudden cardiac death

1. Introduction

Coronary artery anomalies (CAAs) are congenital variations in the origin or course of the coronary arteries. While many are asymptomatic, certain anomalies can lead to significant clinical issues, including myocardial ischemia, arrhythmias, and sudden cardiac death. Computed Tomographic Coronary Angiography (CTCA) has become a pivotal non - invasive imaging modality for diagnosing and evaluating these anomalies.

2. Case Presentation

A 73 - year - old Indian male with no significant past medical history was evaluated in cardiology OPD with complaints of breathlessness on exertion and intermittent chest pain. Both breathlessness and chest pain were exertional, pain was located in the center of chest and non - radiating. The patient was a retired bank employee and denied symptoms of palpitations, dizziness, or syncope with exertion. He underwent a Treadmill Test (TMT) which showed positive results. He denied personal history of heart problems or family history of premature coronary artery disease, inherited arrhythmias, or sudden cardiac death.

Electrocardiogram (EKG) showed normal sinus rhythm with no ECG changes suggestive of ischemia and blood reports showed normal serum cardiac markers. Transthoracic echocardiogram showed normal ejection fraction of 60% - 65% with no wall motion or valvular abnormalities.

3. Case Findings

He underwent coronary computed tomography (CT) angiogram (CCTA) to evaluate the coronary arteries for the presence of Coronary Artery Disease (CAD) which revealed the following findings:

1) Dominant right coronary artery (RCA) with mixed plaques causing 70 - 80 % luminal narrowing proximally and complete thrombosis distally.
2) Anomalous origin of Left Main Coronary Artery (LMCA) from proximal RCA. LMCA had a malignant course between aorta and pulmonary artery.
3) Retrograde filling of PDA through LAD
4) Presence of a Ramus intermedius branch
5) Ostial narrowing of Left Circumflex Artery (LCx)

Results of CT Coronary Angiography: ***CAD - RADS: 5/P4/E/HRP with Double Vessel Disease, Anomalous origin of*** LMCA from RCA with a malignant course between aorta and pulmonary artery.

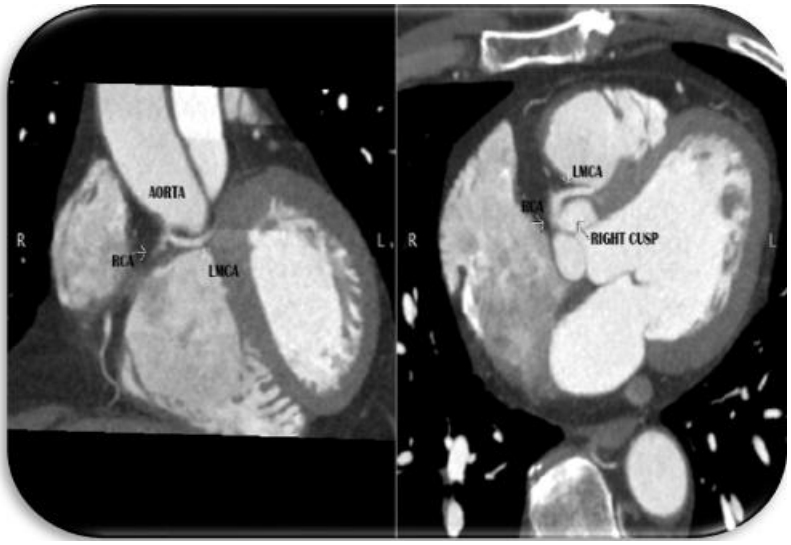
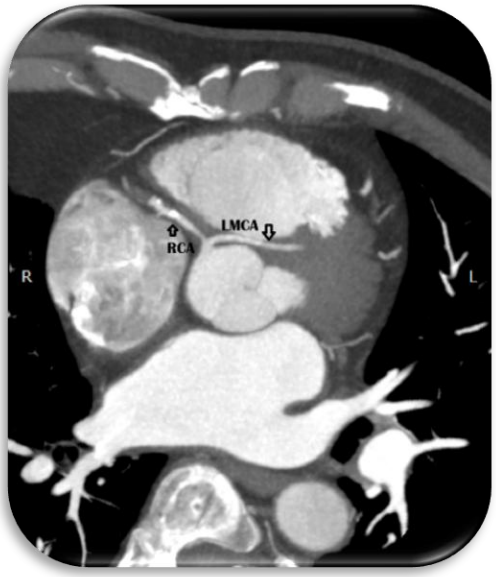
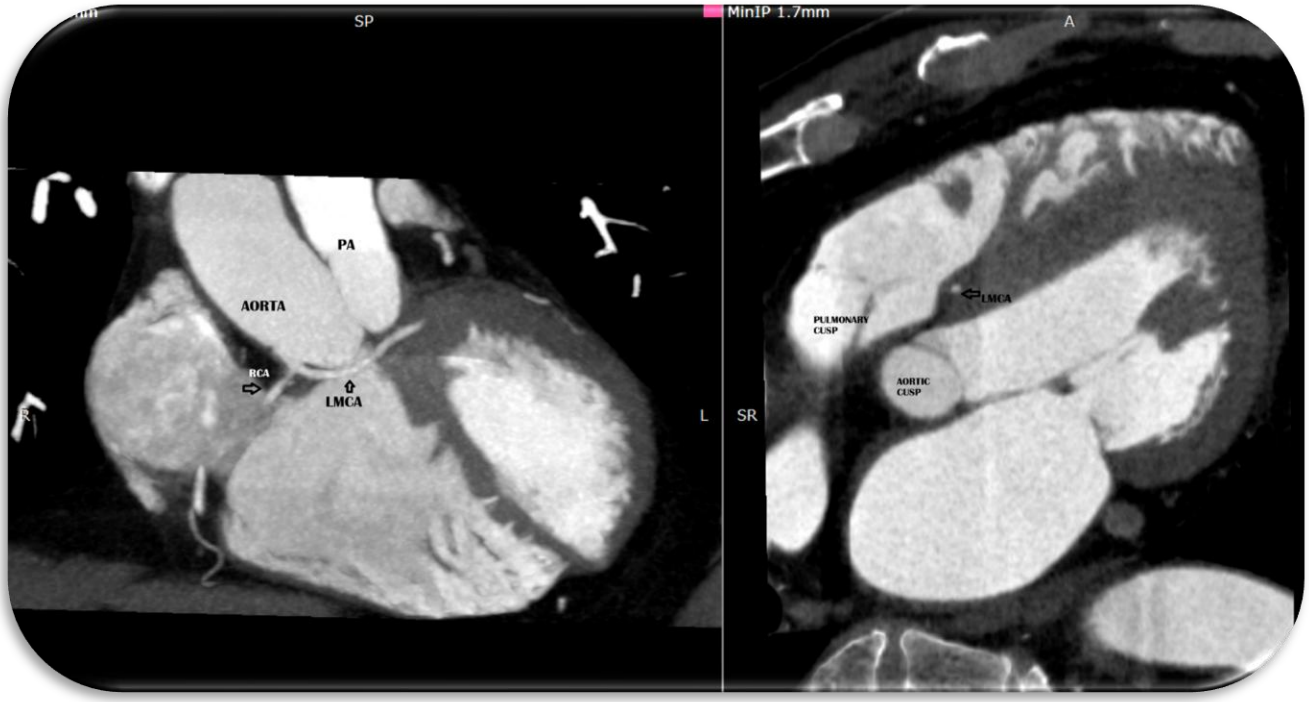


Figure 1: Multiplanar reconstruction of CT showing Anomalous origin of Left Main Coronary Artery (LMCA) from proximal portion of Right Coronary Artery (RCA)





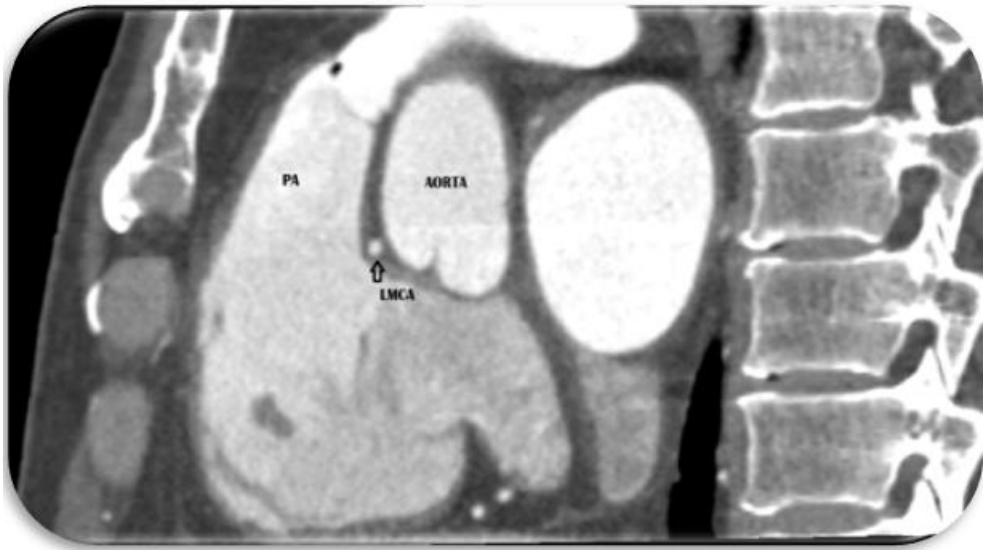


Figure 2: Multiplanar reconstruction of CT showing Inter-arterial course of Left Main Coronary Artery (LMCA) between the Aorta and the Pulmonary Artery (PA)

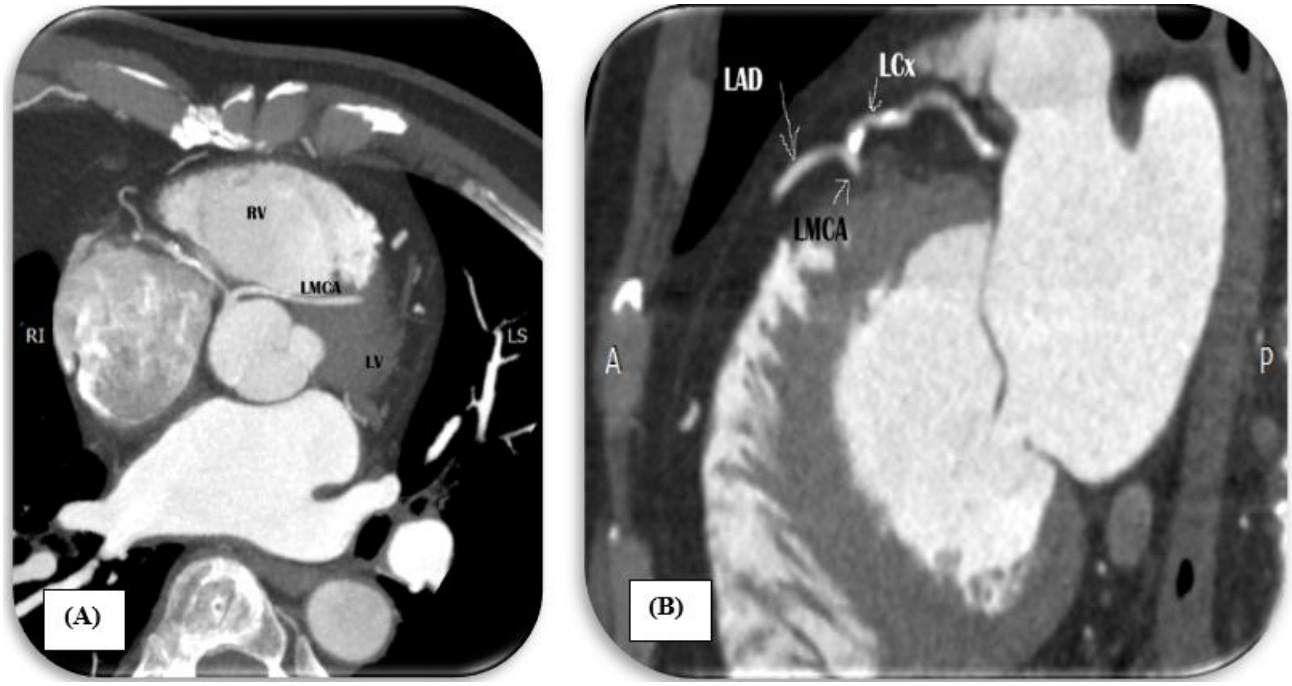


Figure 3: Multiplanar reconstruction of CT showing (A) A long Trans-septal course of Left Main Coronary Artery (LMCA) between the Right Ventricle (RV) and Left Ventricle (LV) and (B). Late Bifurcation of LMCA into Left Anterior Descending (LAD) and Left Circumflex Artery (LCx)

Post CT - Coronary Angiography, on the basis of CAD - RADS 2.0 Score, patient was advised further evaluation with Invasive Coronary Angiography (CAG) which confirmed the above findings.

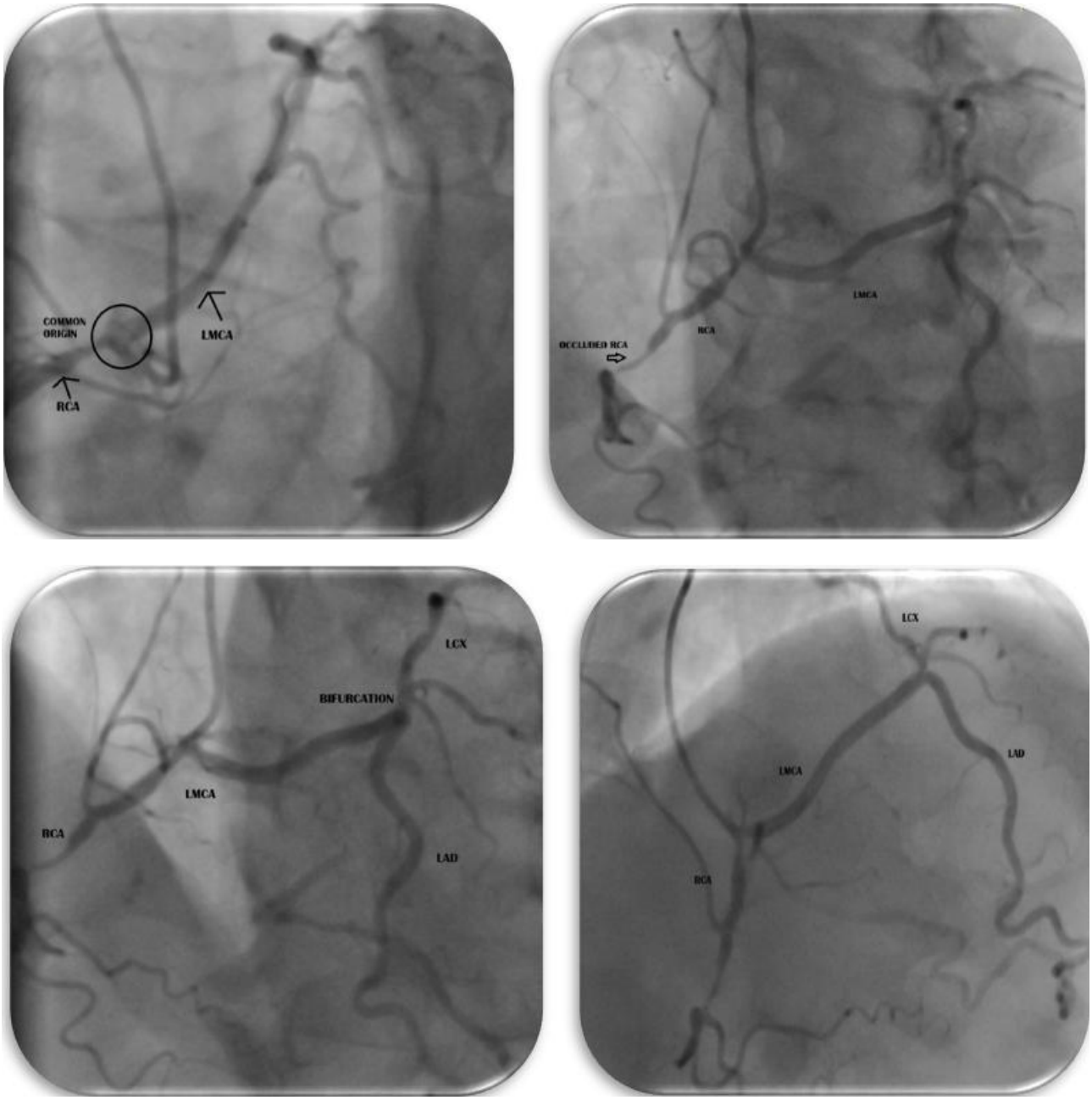


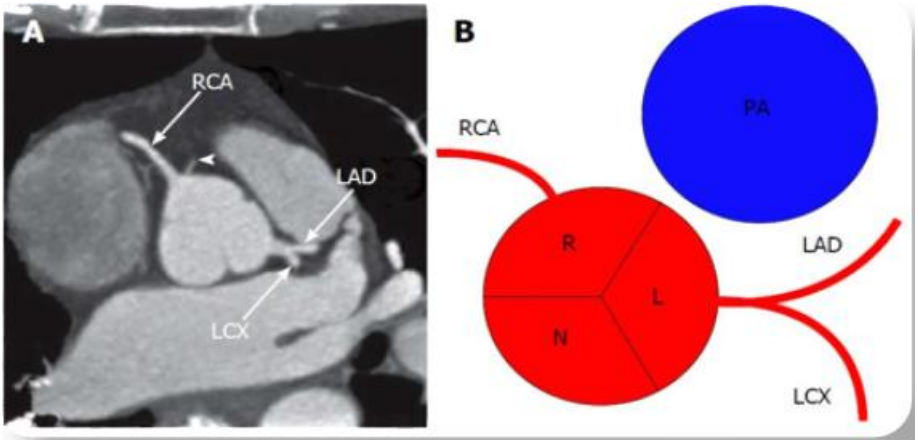
Figure 4: Invasive Coronary Angiography (ICA) images showing common origin of RCA and LMCA, occluded RCA segment and late bifurcation of LMCA into LAD and LCx

4. Discussion

Normal Coronary Anatomy

In typical anatomy, the coronary arteries arise from the aortic sinuses:

- **Left Main Coronary Artery (LMCA):** Originates from the left sinus of Valsalva, bifurcating into the LAD and Left Circumflex (LCx) arteries.
- **Right Coronary Artery (RCA):** Originates from the right sinus of Valsalva, supplying the right ventricle and parts of the posterior left ventricle.



Normal Coronary Anatomy: (A). A Computed Tomography Scan showing normal origin of coronary arteries (B). Diagrammatic representation

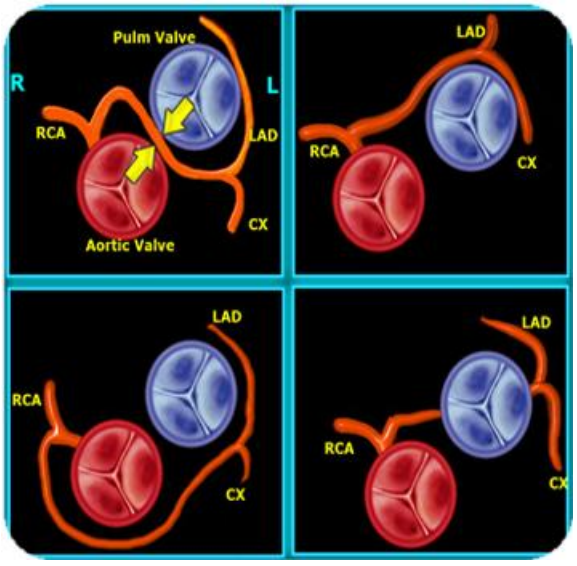
Anomalous Left Coronary Artery: Malignant Course

Anomalous origin of the LCA can occur in several forms:

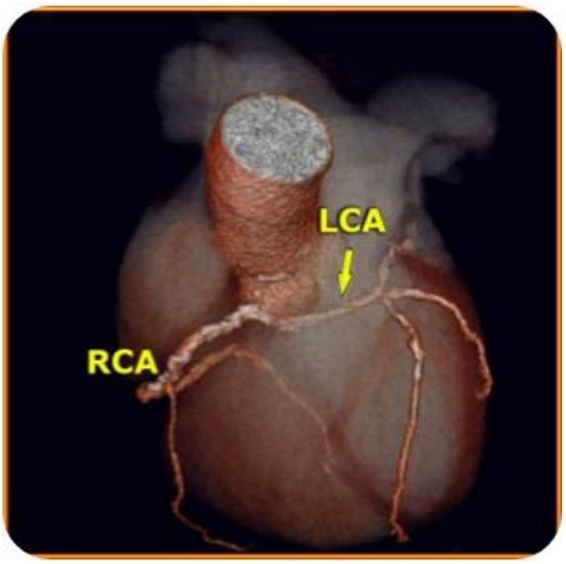
- **Originating from the RCA or right sinus of Valsalva**: This is the most concerning variant, especially when the artery takes an interarterial course between the aorta and pulmonary artery.
- **Originating from the pulmonary artery**: A rare and fatal anomaly leading to myocardial ischemia and sudden death if untreated.
- **Single coronary artery**: A condition where both the LAD and LCx arise from a single ostium, increasing the risk of ischemia.

The inter - arterial course is particularly dangerous due to the potential for compression between the aorta and pulmonary artery during systole, leading to myocardial ischemia and arrhythmias.

Anatomical variants of origin and course of coronary arteries



Anatomical variants of origin and course of coronary arteries



3D Volume Rendered Image showing Anomalous origin of LCA from Right Aortic Cusp

Pathophysiology

The malignant course of the LCA can lead to:

- **Interarterial compression**: The artery is compressed between the expanding aorta and pulmonary artery during systole, reducing blood flow.
- **Acute angle takeoff**: An abnormal origin angle can cause a slit - like orifice, further compromising blood flow.
- **Intramural segment**: The artery may travel within the aortic wall, increasing resistance and susceptibility to ischemia.

These factors can result in myocardial ischemia, arrhythmias, syncope, and sudden cardiac arrest, particularly during physical exertion.

Clinical Presentation

Symptoms may include:

- Exertional chest pain
- Syncope or presyncope
- Palpitations
- Sudden cardiac arrest, especially in young athletes
- Asymptomatic cases detected incidentally

Diagnosis

Imaging techniques are crucial for diagnosis:

- **Coronary CT Angiography (CTA):** Provides detailed visualization of anomalous coronary anatomy. It can be further supported by a detailed CT - FFR study which allows for the assessment of the functional impairments caused by these anomalies without the need for invasive procedures.
- **MRI and Echocardiography**: Useful adjuncts, especially in pediatric patients.
- **Coronary Angiography**: Invasive but can assess hemodynamic significance.
- **Stress Testing**: May reveal ischemia but carries risk in known malignant courses.

Management

Management strategies include:

1) **Conservative Approach**: For asymptomatic, low - risk patients—activity restriction and regular monitoring.
2) **Surgical Intervention**: For symptomatic or high - risk patients:
   - **Unroofing Procedure**: Removing the intramural segment to allow unobstructed flow.
   - **Coronary Reimplantation**: Repositioning the anomalous artery to its appropriate sinus.
   - **Coronary Artery Bypass Grafting (CABG):** Used if direct repair is not feasible.

The choice of procedure depends on the anatomy, patient age, and surgical expertise.

Prognosis

Early detection and appropriate management significantly improve outcomes. Surgical correction reduces the risk of sudden cardiac death and enhances quality of life. However, undiagnosed cases, especially in young individuals with high physical activity levels, remain at risk for catastrophic events.

5. Conclusion

Anomalous origin of the LMCA with a malignant course is a serious congenital anomaly that warrants heightened clinical awareness due to its potential lethality. With advances in imaging and surgical techniques, early diagnosis and appropriate intervention can be life - saving. Clinicians should maintain a high index of suspicion in young individuals with unexplained exertional symptoms, especially athletes, and ensure thorough cardiac evaluation.

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