Chapter 35
A Rare Case of Adult Type ALCAPA Syndrome

Bahram Mohebbi

Abstract A 43-year-old male patient complained with atypical chest pain and dyspnea on exertion since six months ago. In heart physical examination, S1, S2, and III/VI systolic murmur in left sternal border was detected. In transthoracic echocardiography, mild LV enlargement with severe systolic dysfunction, EF=30% was reported. Patient was candidate for selective coronary angiography. Based on coronary angiography, Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) syndrome was confirmed and referred to cardiac surgeon to manage his condition.

History and Clinical Presentation

This case report described a 43-year-old male patient presented with atypical chest pain and dyspnea on exertion with slight limitation of physical activity, (New York Heart Association [NYHA] class II) since 6 months ago. The patient's vital signs were blood pressure 120/70 mmHg, pulse 80 beats/min with a regular rhythm. He did not report any cardiovascular risk factor. In heart physical examination, S1, S2, and III/VI systolic murmur in the left sternal border was detected.

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Para-Clinic Assessment

In transthoracic echocardiography, mild LV enlargement with severe systolic dysfunction, EF = 30% was reported. The patient underwent coronary angiography. Access was made through the right radial artery and due to the inability to engage the left coronary system, nonselective aortic root injection revealed only right coronary artery (RCA) origination from aorta which filled the left coronary system retrogradely (Video 35.1).

In selective RCA angiography, RCA was dominant and huge which retrogradely filled the left coronary system and drained to the pulmonary artery(PA) (Fig. 35.1) (Video 35.2). Actually, the left coronary system was originated from PA with retrogradely filled via RCA with good distal runoff.

Management

Because of enhancing survival rate due to performing surgery among pediatric ALCAPA patients, in this case, assessing the risk/benefit for surgery; finally, surgical correction was considered to the patient which was performed successfully.

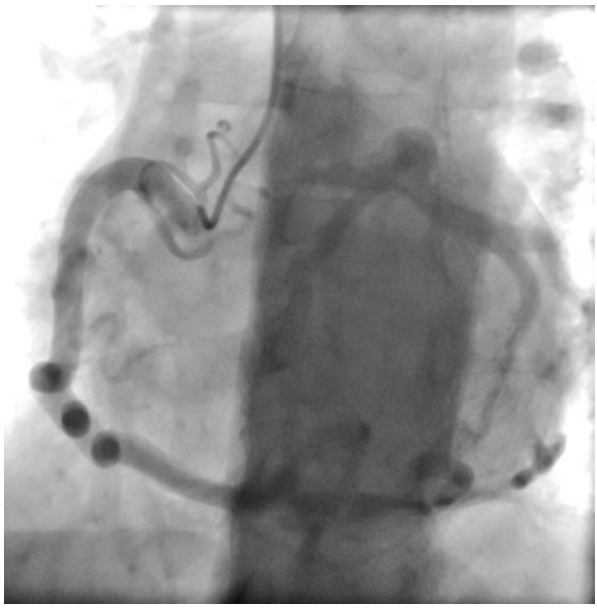

Fig. 35.1 Selective RCA angiography. RCA was dominant and huge which retrogradely filled left coronary system and drained to PA

Conclusion

Coronary artery anomalies consist of several variants with diverse cardiovascular manifestations. In order to categorize each anomaly, distinguishing any condition of anatomic pattern and clinical significance to understand the prognosis is crucial. ALCAPA syndrome is an uncommon diagnosis. Generally, it is very rare for ALCAPA syndrome to be diagnosed in adulthood, because if they did not receive on-time treatment in childhood, this malformation can lead to death [1]. Due to low pulmonary artery pressure, flow in the left coronary artery reverses and drains to the pulmonary artery instead of supplying the myocardium completely. So there is a steal phenomenon due to this left-to-right shunting [2]. The final decision in patients with ALCAPA syndrome is surgery. There are some surgical techniques as left main (LM) coronary artery reimplantation, Takeuchi procedure, left coronary artery ligation, and left coronary artery ligation with concomitant coronary artery bypass grafting [3].

References

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