Acute coronary syndrome caused by anomalous origin of the right coronary artery from the left sinus of Valsalva

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Summary
An anomalous origin of right coronary artery from the left sinus of Valsalva is a rare congenital anomaly of the coronary arteries. We describe a 40-year-old male hypertensive and hyperlipidemic patient who presented with typical anginal pain with normal ECG and echocardiogram and abnormal exercise ECG Test. Coronary angiography showed an anomalous origin of the right coronary artery from the left sinus of Valsalva, an anomaly that has been associated with angina, myocardial infarction and sudden cardiac death. The clinical profile of this congenital anomaly is also reviewed.

Key-words: Congenital coronary anomaly, Anomalous origin of right coronary artery.

Résumé
Une origine anormale d’artère coronaire de côté droite à partir du sinus de valsalva du côté gauche est une anomalie congénitale des artères coronaires rare. Nous présentons le cas d’un patient de sexe masculin âgé de 40 ans hypertendu et hyperlipidémique qui s’est présenté atteint d’une douleur anginale typique avec ECG normal et échocardiogramme et exercice abnormal d’examen ECG. L’angiographie coronaire a montré une origine anormale de coronaires du côté droit à partir du sinus de valsalva du côté gauche, une anomalie qui a été liée avec infarctus du myocarde angina et la mort à travers une crise cardiaque. On a également fait le bilan du profil clinique de cette anomalie congénitale.

Introduction
The incidence of congenital coronary anomalies varies from 0.2% to 1.2% of the general population1. The majority of the anomalies are benign but few are associated with serious outcomes.

We describe the case of a patient who presented with chest pain, and coronary angiography revealed an anomalous origin of the right coronary artery from the left sinus of Valsalva. We discuss the clinical significance of this anomaly and review the literature for the latest update and management.

Case presentation
A 40-year-old male teacher with history of hypertension and hyperlipidemia presented to cardiology clinic/Asfer Central Hospital complaining of exertional retro-sternal chest pain relieved by rest and associated with dyspnea and sweating for the last one year. Physical examination revealed average built male, his BP was 160/95 mmHg, heart rate was 80 beats/minute, regular rhythm with normal volume and character and no radio-femoral delay.

His JVP was not elevated, and auscultation revealed normal first and second heart sounds with additional fourth heart sound.

The rest of his systemic examination was within normal limits. Routine laboratory work was normal except mildly elevated cholesterol level (total cholesterol of 225 mg/dl and LDL of 110 mg/dl).

Exercise treadmill testing (ETT) was reported to be positive for myocardial ischemia, with chest pain and ST-segment changes at stage four of Bruce protocol.

Echocardiography showed normal study. The patient was referred for cardiac catheterization where selective coronary catheterization of the left coronary system showed non-obstructive coronary artery disease, and showed an anomalous origin of the right coronary artery from the left coronary sinus (Fig. 1). The patient was started on B.Blocker and his anginal symptoms resolved during follow-up.

Discussion
Anomalous origin of right coronary artery from the left sinus of Valsalva is a rare congenital anomaly of the coronary arteries.

It is first described in 1948 by White and Edwards2. The prevalence of this congenital anomaly varies according to the race, where it is approaching only 0.026% in autopsy studies of white population.3 The angiographic prevalence in Japanese population is estimated to be 0.25%.4

The clinical significance of this rare anomaly is not clear and ambiguous. Varying from no impact on life expectancy...
to high incidence of Sudden Cardiac Death.\(^{(5)}\) Full-spectrum of clinical presentation including origin, myocardial infarction and even sudden death have been described.\(^{(6)}\)

In 1992, Taylor and co-authors studied 52 patients with anomalous origin of the right coronary artery and reported 25% incidence of sudden death.\(^{(5)}\)

The pathophysiology involved is still not clear, theories proposed to explain it include; mechanical compression of the anomalous artery between the great vessels mainly the aorta and pulmonary artery as it passes in between, the oblique angle at the take off of the anomalous artery from the aorta and subsequently the slit-like orifice in the aortic wall,\(^{(5)}\) and finally the vulnerability of the proximal segment of the anomalous artery to spasm was also reported.\(^{(5)}\)

In an attempt to find out the clinical or pathologic variables that can predict sudden death, Taylor found only the age of 30 or older to be the only prediction of lower incidence of sudden cardiac death.\(^{(7)}\) Recently MRI was advocated to be a useful non-invasive test for such an anomaly, where the course of the artery can be delineated.\(^{(8)}\)

Treatment of such an anomaly is also controversial. Surgical options include many techniques but no long-term benefits have been established. Some of these techniques include: translocation of the anomalous artery to the aorta\(^{(9)}\), bypass grafting of the anomalous right coronary artery and ligation of the native proximal portion\(^{(10)}\), excision of the common wall between the right coronary artery and the aortic wall\(^{(11)}\).

Conservative non-surgical treatment has also been advocated using B-blocker therapy with favorable response, in Japanese experience with 56 patients with this anomaly treated with B-blocker therapy, no mortality was reported directly related to the anomaly during 5-year follow up period.\(^{(12)}\)

In our case the patient was started on beta blockers (oral Metoprolol 50 mg twice daily), which was effective in resolving the anginal symptoms.

References
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