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Case Report

Acute Coronary Syndrome Revealing Coronary Birth Abnormality in A Patient with an Arrhythmogenic Dysplasia of the Right Ventricle: Case Report

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Abstract: Abnormal coronary artery birth is a congenital abnormality that remains rare. In some cases, congenital coronary abnormalities may be associated with the development of chest pain, syncope or sudden cardiac death. Our patient is 65 years old with a chronic smoking cardiovascular risk factor and as a history a right ventricular arrhythmogenic dysplasia discovered for 7 years. Our patient was admitted for acute Coronary Syndrome NSTEMI , a coronary angiogram was performed showing coronary arteries without significant stenosis. However, the right coronary artery had an abnormal birth in the left sinus more precisely in the left main. We are reporting the case of a patient who presented for acute coronary syndrome as revealed by typical anginal chest pain, highlighting that he is being followed for arrhythmogenic dysplasia of the right ventricle, with an abnormal birth of the right coronary artery at of the left main. Several studies have been conducted to establish the relationship between the patient's symptoms, his history and this angiographic finding

Keywords: Acute coronary syndrome, Coronary birth abnormality, Arrhythmogenic dysplasia of the right ventricle.

INTRODUCTION:

Abnormal coronary artery birth is a congenital abnormality that remains rare. In some cases, congenital coronary abnormalities may be associated with the development of chest pain, syncope or sudden cardiac death (Cheitlin, M. D. *et al.*, 1974; & Taylor, A. J. *et al.*, 1992). We are reporting the case of a patient who presented for acute coronary syndrome as revealed by typical anginal chest pain, highlighting that he is being followed for arrhythmogenic dysplasia of the right ventricle, with an abnormal birth of the right coronary artery at of the left main. Several studies have been conducted to establish the relationship between the patient's symptoms, his history and this angiographic finding.

RESULTS:

Our patient is 65 years old with a chronic smoking cardiovascular risk factor and as a history a right ventricular arrhythmogenic dysplasia discovered for 7 years under beta-blockers and having for several months angina Class II of the CCS. Our patient was admitted to the ER for retrosternal chest pain radiating to the upper left limb. He had a good hemodynamic and respiratory condition with a physical examination without abnormalities. An electrocardiogram was made recording a regular sinus rhythm, a right branch block with secondary repolarization disorders and a planing of the lower R-wave. A determination of the ultrasensitive troponins was made at the intake with a high resulting rate, as well as the CK-MB. Other biological tests, including serum electrolytes, were normal, as was chest radiography. As a result, the diagnosis of Acute Coronary Syndrome NSTEMI was retained and the patient was hospitalized in the ICU. A transthoracic echocardiography was performed that detected a left ventricle with size and function maintained, correct global and segmental kinetics and a normal ejection fraction, in return, the right ventricle was not dilated, hypertrophied with akinesia of its lateral wall suggestive of right ventricular dysplasia. The patient was treated with platelet antiblockers; Aspirin, Clopidogrel, intravenous heparin, beta blockers, conversion enzyme inhibitors, and statins. A coronary angiogram was performed showing coronary arteries without significant stenosis. However, the right coronary artery had an abnormal birth in the left sinus more precisely in the left main . Acute aortic syndromes and pulmonary embolism were excluded. Thus, the possibility of an abnormal path of this artery particularly in inter aorto pulmonary was suspected. A thoracic coronary scan was therefore requested which

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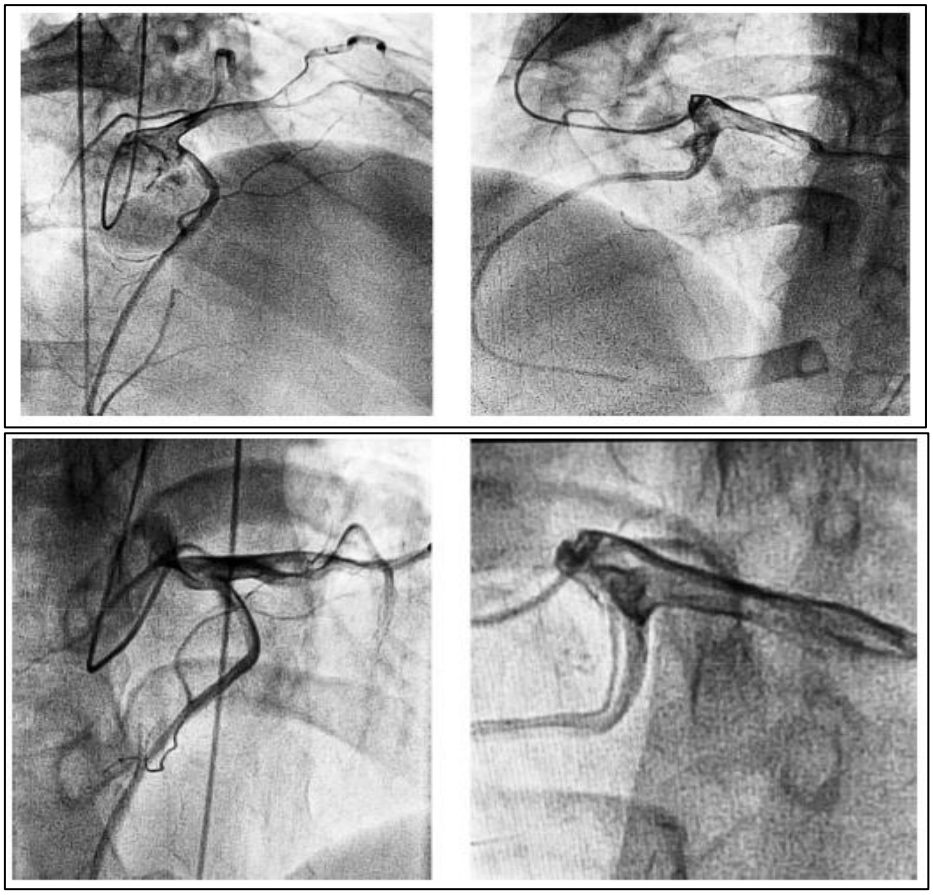
was unfortunately not carried out because of the patient's material difficulties.



Images Showing the Abnormal Birth of the Right Coronary Artery from the Left Main

DISCUSSION:

Coronary artery birth abnormalities have intrigued clinicians, anatomists and physiologists for many years. Since the advent of coronary angiography in the 1960s, this segment of systemic circulation has become a common subject of exploration (Angelini, P. 1989). After hypertrophic cardiomyopathy, abnormal coronary artery births are the second leading cause of sudden death in young athletes. In a recent study, birth abnormalities in one or more coronary arteries caused the death of up to 19% of young athletes who died during or shortly after intense physical activity (Maron, B. J. *et al.*, 1996). The abnormal birth of coronary arteries may be asymptomatic or prove to be extreme by sudden death especially if the patient is already being followed for a rhythm disorder as is the case in our patient with arrhythmogenic ventricular dysplasia. The embryological explanation for the abnormal birth of coronaries was explained by Angelini, P. (1989). Initially, the vascular sinusoids develops within the embryonic myocardium before its definitive compaction. At the same time, epicardial blood islets coalesce into a rudimentary plexus. After a long period of controversy over the origin of coronary ostia, it is now known that they reach the aorta centripetally towards each of the corresponding Valsalva sinuses. After regression of the sinusoids and connection of the epicardial plexus with the coronary sinus, the definitive network is formed (Majesky, M. W. 2004; & Olivey, H. E. *et al.*, 2004). There is no precise definition that a distribution or route should be considered a variant rather than an anomaly when it is present in at least 1 % of the population (Levin, D. C. *et al.*, 1978). Thus, the presence of three coronary ostias can be considered normal. The initial pathof the coronary arteries are usually perpendicular to the sinus but a wide variety of orientations exist up to the tangential path considered abnormal. Finally, the distribution of epicardial arteries and branches of division is extremely varied and, again, defining normality is difficult (Raimondi, F., & Bonnet, D. 2016). Our case was admitted for Acute Coronary Syndrome with coronary angiography detecting significant arteries without stenosis, so an inter-arterial path of the right coronary artery is the most plausible explanation of the symptoms the patient presents. A higher risk of death was associated with this type of abnormality explained by the compression of the right coronary artery impacted between the aorta and the pulmonary trunk during the effort resulting in ischemia which can probably explain the angina but can also manifest by a syncope, congestive heart

failure, arrhythmia and sudden death (Brandt III, B. *et al.,* 1983; Frescura, C. *et al.,* 1998; Isner, J. M. *et al.,* 1984; Kimbiris, D. E. M. E. T. R. I. O. S. *et al.,* 1978; & Reul, R. M. *et al.,* 2002) especially if the patient is already being followed for arrhythmogenic dysplasia of the right ventricle, further aggravating the rhythmic prognosis. Right ventricular dysplasia is a genetic cardiomyopathy whose transmission is usually autosomal dominant at which the heart muscle cells are gradually replaced by fat and fibrous tissue. The disease concerns all the myocardium but especially the right ventricle. It exposes in the same way as the abnormal births of coronaries to disorders of the rhythm most often ventricular with sometimes a sudden death specially described in young athletes for these two entities. This phenomenon is explained in right ventricular arrhythmogenic dysplasia by the impact of fibro-adipose tissue on the

electrical equilibrium of the heart resulting rhythm disturbances through the development of the intercellular junctions characterising the disease (Basso, C. *et al.,* 2009; & Saffitz, J.E. 2005) and at the level of abnormal birth by either a spasm or a compression of an artery following its abnormal path which is the most likely theory in our patient (Swaminath, D. *et al.,* 2013). The birth of the right coronary artery to the left coronary sinus is six times more common than the opposite situation of the left coronary artery of the right coronary sinus. Therefore, although surgical repair is recommended for all patients with the left coronary artery from the right coronary sinus, this is not the case with the right coronary artery from the left coronary sinus, where it is advisable to select only a subgroup of patients (Warnes, C. A. *et al.,* 2008). Surgical options for the anomalous coronary origins of the contralateral sinus are multiple to type of bypass, re-implantation of the anomalous vessel in its appropriate sinus (Peñalver, J. M. *et al.,* 2012). Non-surgical strategies include the use of beta-blockers and avoidance of participation in all competitive sports for patients (Warnes, C. A. *et al.,* 2008).

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