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LETTER TO THE EDITOR

Fistulous coronary artery aneurysms: Further insights into mechanistic and clinical implications

Fístulas em aneurismas das artérias coronárias: novos dados sobre implicações mecanicistas e clínicas

Coronary artery fistula (CAF) is a rare vascular phenomenon primarily characterized by a fistulous connection between major coronary arteries and low-pressure cardiac chambers.[1–3] Clinically, this phenomenon might present with a variety of clinical manifestations including cardiac volume overload, myocardial ischemia and coronary aneurysm formation, etc., which might warrant percutaneous or surgical management strategies in certain settings.[1–3] The recently article by Briosa e Gala, et al. described a case of giant right coronary artery (RCA) aneurysm with a fistulous drainage into the coronary sinus in a middle-aged female patient.[1] Accordingly, we would like to comment on this interesting case and further implications of fistulous coronary aneurysms in clinical practice.

First, CAFs arise principally as congenital diseases in the overwhelming majority of cases, most often in the form of a fistulous connection between the RCA and right ventricle.[2,3] However, acquired forms due to trauma or coronary interventions, although rare, have also been reported.[2] The present case might possibly have a congenital origin,[1] nevertheless, we wonder if there is further information in the patient's history that might suggest there are acquired causes (if any) for the CAF formation.

Importantly, CAF, whether congenital or acquired, may be regarded as the primary disease in most patients with fistulous coronary aneurysms, potentially suggesting CAF flow as the most important determinant of clinical outcomes.[2,3] In other terms, associated coronary aneurysm formation in these patients mostly arises as a secondary phenomenon that emerges successively and progresses over time. Terminologically, these CAFs may be termed as 'primary CAFs' (with or without coronary dilatation or aneurysm). Of note, primary CAFs involving the relatively distal coronary segments generally do not lead to aneurysm formation, and mostly presents with relatively small[3] and diffuse dilatation of the associated coronary artery. In patients with primary CAF, coronary dilatation and aneurysm formation, besides potentially having a congenital background to some degree,[3,4] may also arise as part of maladaptive changes in response to coronary steal phenomenon, and; hence, may be indicative of myocardial ischemia (at rest or during stress) in these patients. Accordingly, we wonder about signs and symptoms of myocardial ischemia on electrocardiogram or stress testing modalities (if any) in this patient.

Second, CAF can occasionally emerge as a potential complication (therefore a secondary phenomenon) of pre-existing coronary aneurysms[4] possibly due to the consequent mural erosion involving the aneurysmal sac and neighboring structures. Terminologically, these CAFs might be classified as secondary CAF, and may be encountered only in a small portion of fistulous coronary aneurysms. Importantly, secondary CAFs generally complicate the giant aneurysms (renowned for their unfavorable clinical outcomes) potentially associated with atherosclerosis, Kawasaki disease (KD, Takayasu arteritis as well as connective tissue diseases including Marfan syndrome (each presenting with diverse histopathological characteristics).[4] Of note, fistulous coronary aneurysms in this context can be located elsewhere along the coronary artery with a particular predilection for proximal coronary segments in certain conditions including KD.[4,5] Prognostically, emerging secondary CAFs within the pre-existing aneurysmal sacs may be predictors of more serious complications including imminent aneurysm rupture, etc., and hence might require urgent management strategies (primarily targeting the aneurysmal sac),[4] regardless of the severity of CAF flow and symptomatology. Within this context, the giant nature of the aneurysm in the patient[1] potentially warrants further clinical investigation. Accordingly, we wonder whether the patient had a history of systemic vasculitis or findings of systemic inflammation (inflammation markers, etc.) or signs of existing connective tissue disease? We also would like to have information regarding the histopathological findings of the excised material.

Finally, there is no uniformly agreed absolute threshold diameter to accurately define giant coronary aneurysms.[4,6] Threshold values of >20 mm and >50 mm have been previously suggested in the context of congenital coronary aneurysms.[4,6] However, more objective calculations including the ratio of aneurysm/reference segment diameter have also been recommended for the absolute definition of giant coronary aneurysms (a ratio of >4 signifies a giant aneurysm).[4,6] Moreover, calculation of 'Z score' (standard deviation from the means) has been the preferred strategy owing to its well-known potential to adjust for confounding

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factors, including body surface area, gender, etc.[5,6] In the setting of KD, a Z score value of >10 was previously suggested to define giant coronary aneurysms.[5,6] We therefore wonder about the values of aneurysm/reference segment diameter ratio and Z score in the patient.

In conclusion, CAF can be predominantly considered as the primary disease (and mostly congenital) in patients with fistulous coronary aneurysm potentially suggesting CAF flow as the substantial determinant of clinical outcomes and subsequent management strategies. On the other hand, CAF formation may occasionally emerges as a potential complication (secondary CAF) of the pre-existing coronary aneurysms (particularly the giant ones), and usually warrants urgent management strategies due to the impending risk of catastrophic complications (aneurysm rupture, etc.). In the setting of fistulous coronary aneurysms, it seems crucial to differentiate between these two types of CAFs largely through evaluation of coronary aneurysm characteristics (size, coronary segmental location, etc.), history and clinical findings to improve risk-stratification and management of these aneurysms. However, further implications of fistulous coronary aneurysms still need to be fully established.

Conflicts of interest

The authors have no conflicts of interest to declare.

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