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EDITORIAL COMMENT

Be mindful when evaluating a patient with chest pain and pulmonary hypertension

Esteja alerta quando estiver a avaliar um doente com dor torácica e hipertensão pulmonar

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The case[1] presented in this issue of the Portuguese Journal of Cardiology illustrates the clinical approach to a young, female patient with chest pain and pulmonary hypertension (PH). While this case focuses on a specific cause of chest pain, the reader should be reminded about some crucial aspects when evaluating this symptom in PH patients.

First and foremost, resist the classic temptation to attribute angina and angina-like symptoms to the increased and unmatched metabolic demands of the hypertrophied and overloaded right ventricle. The reasons are twofold: 1) PH is a devasting disease, with more than 25% of deaths cases of sudden cardiac death (SCD)[2]; 2) some of the causes of SCD can be easily recognized and are, therefore, preventable.

In this regard, the causes of SCD in PH can be divided in two major categories: arrhythmic and non-arrhythmic. While it is known that ventricular arrhythmias in PH are predominantly described in congenital heart disease patients, preventing this dreadful complication is not an easy task. On the other hand, non-arrhythmic causes such as pulmonary artery (PA) dissection, PA rupture, massive hemoptysis and left main (LM) compression syndrome are mainly associated with dilatation of the PA. Amongst these, LM compression syndrome can be identified early with computerized tomography coronary angiogram, which gives us a chance to correct it and potentially avoid SCD.

As the name implies, after LM compression syndrome is characterized by an extrinsic compression of the LM coronary artery by a dilated PA and has been increasingly recognized as a cause of angina in PH patients (estimated prevalence 15 to 30%).[3] Nevertheless, be mindful when considering treatment for PH patients with possible LM compression syndrome. After all, and especially when dealing with a young population, you will consider stenting the LM or PA surgical plasty for your patient. If you do, remind yourself of the following: 1) LM stents are not complication-free (e.g., stent thrombosis or restenosis); 2) surgery in PH is not low risk; and 3) coronary blood flow and subendocardial perfusion are diastolic events while LM compression from an enlarged PA is a systolic event – meaning that not all lesions will lead to ischemia. As such, LM compression syndrome patients with atypical symptoms and no evidence of rest ischemia should be further tested to assess the physiological significance of the stenosis. Exercise stress testing is relatively contraindicated in patients with severe PH, and vasodilator stress nuclear testing may miss global ischemia in patients with LM disease. Thus, stress cardiac magnetic resonance, instantaneous wave-free ratio and fractional flow reserve could be better suited to assessing stenosis significance in this setting,[4,5] and drastically change patient management.

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As a final reminder, the reader should never forget atherosclerotic coronary artery disease, especially in a PH patient with several atherosclerotic risk factors and typical angina. Ultimately, it would be shameful if we missed this one.

Conflicts of interest

The author has no conflicts of interest to declare.

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