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Small vessel disease in ARVD and HCM

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S mall vessels in the heart are distal coronary arteries with a diameter between 50 and 200 microns. The same histologic pattern is found in two inherited cardiomyopathies ARVD and HCM. The typical pattern is an increase in the thickness of the leiomyocytes in the media of these vessels. There is also a reorientation of these leiomyocytes which are no longer parallel to the vessel but perpendicular. In addition, this anomaly seems independent of the surrounding tissue which can be dysplastic or not in ARVD. This histological structure is the basis of "atypical" chest pain which is the main clinical sign of the small vessel disease. Atypical means that they are not effort related as in common coronary artery disease. Their occurrence is unpredictable and duration variable from seconds to hours as well as their intensity. They occur more frequently in middle age women triggering multiple investigations to assess coronary vessels which are all negative. The histological structure of the distal coronary vessel may explain chest pain by spasm produced by the contraction of leiomyocytes leading to ischemia. An impressive case of ARVD was a 30 years old woman with frequent atypical chest pain even awaking her at night. Standard coronarography was normal. However, an ergonovine test triggered her typical episode of chest pain and a spasm of 1.5 cm was observed on the middle of the main descending artery. Pain and spasm disappeared after vasodilator injection. In her case ARVD was diagnosed by atrial arrhythmias (flutter) and extrasystoles with a left bundle branch block. ARVD was confirmed by ventriculography of the right ventricle. However, a systematic histologic study of the right ventricle of 82 patients who died of a non-cardiac cause in a general hospital of Paris showed that a quiescent form of ARVD (no arrhythmias) was observed in 3,7% of cases (Fontaine Editorial AJC 2014). Therefore, it can be that atypical chest pain commonly observed in middle age women are in fact the expression of a concealed form of ARVD. The presence of a similar histologic pattern and atypical chest pain in ARVD and HCM suggests a genetic mechanism which is presently unknown. This new concept needs further studies.

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