Primary cardiac lymphoma causing coronary vasospasm

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A 63-year-old woman with no significant medical history or risk factors presented with an acute coronary syndrome. It was her first episode of angina pectoris (AP). ECG showed ST-segment elevations in II, III, and aVF, and cardiac biomarkers were negative. Coronary angiography revealed a severe stenosis in the right coronary artery (RCA) (Panel A), which disappeared after intracoronary nitroglycerin infusion. Subsequent coronary vasomotility testing with the intracoronary infusion of acetylcholine induced diffuse coronary vasospasm, reproducing the patient’s initial AP. Despite anti-anginal medication, AP reoccurred almost daily. In order to rule out an ongoing inflammatory myocardial process, cardiac magnetic resonance imaging was performed, which revealed a pericardial mass (56 × 39 mm2) encasing the RCA (Panel B) (Supplementary data online, Movie S1). A biopsy was taken after a mini-thoracotomy, and the histology was consistent with primary cardiac large B-cell lymphoma (Panel C).

Upon the completion of radio chemotherapy, a significant decrease in mass size was found (Supplementary data online, Movie S2) and the patient’s AP disappeared completely. Hence, coronary vasospasm was suspected to be associated with the lymphoma. A possible explanation would be the release of vasoactive agents by tumorous B-cells, inducing coronary vasospasm. Indeed, the majority of lymphoma cells showed immunoreactivity for the vasoactive agent neuropetide-Y (Panel D).

In summary, myocardial ischaemic symptoms in our patient were caused by coronary vasospasms. Most likely, they were induced by neuropetide-Y produced by the lymphoma. The physical neighbourhood of the tumour to the RCA may explain the initial ST-elevations in the inferior leads by neuropetide-Y-induced transient complete vasospastic occlusion.

Supplementary data are available in European Heart Journal — Cardiovascular Imaging online.

Conflict of interest: none declared.

Panel A. Coronary angiography of the RCA before the application of nitroglycerin. Panel B. Cardiac magnetic resonance four-chamber view: round mass encasing the RCA (arrow). Panel C. Histology: sheets of centroblasts showing a basophilic cytoplasm and round nuclei with open chromatin consistent with diffuse large B-cell lymphoma (Giemsa, × 400). Panel D. Immunohistochemical detection of neuropetide-Y in lymphoma cells. The immunoreaction was strong with the peroxidase on the left and the fluorochrome detection method on the mid-picture. Omission of the primary antibody clearly shows the absence of background staining on the right.