Coronary artery disease in systemic sclerosis not clinically apparent: findings from optical coherence tomography

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A 42-year-old female was referred for intermittent effort angina. She had low body mass index, no coronary risk factors and an unremarkable past medical history except for a Raynaud’s phenomenon. Owing to a normal exercise stress test (Panel A), symptoms were initially attributed to anxiety. However, a 24h EKG monitoring revealed diffuse ST-segment depression during physical activity (Panel B). On admission, CT scan showed low-density areas at proximal/mid segments of the left anterior descending artery (LAD), without calcifications (Panel C). At coronary angiography LAD presented with a long, sub-occlusive stenosis (Panels D and E) with collaterals from the right coronary artery. Optical coherence tomography (OCT) demonstrated diffuse intimal-medial thickening of the LAD, a finding suggestive for a fibrotic process involving the vessel (Panel F and Supplementary material online, Video S1). Intravascular ultrasound showed constrictive vessel remodeling (Panel G). Two everolimus-eluting stents were implanted in overlap in the LAD with optimal final result (Supplementary material online, Figures S1 and S2). An OCT pullback of the radial artery documented focal intimal thickening, suggesting different stages of vascular involvement of the medium-small arteries (Panel H). A videocapillaroscopy (Panel I) identified typical features of early scleroderma peripheral microangiopathy, with giant capillaries and haemorrhages. Systemic sclerosis (SSc)-related autoantibodies were still negative.

Systemic sclerosis has a strong macrovascular component with an increased risk of heart attack. Involvement of the medium-small arteries is one of the earliest features of SSc preceding the widespread fibrosis. The present case demonstrates how OCT may orient in the diagnosis and treatment of an uncommon cause of CAD such as SSc not clinically apparent yet.

Supplementary material is available at European Heart Journal online.