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Screening of coronary microvascular dysfunction in cardiomyopathy of myotonic muscular dystrophy type 1

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A 52-year-old, actively smoking and overweight man was referred for invasive coronary angiography (ICA). He had been suffering from type 1 myotonic muscular dystrophy (DM1) for 30 years with discrete motor disorders and dyspnoea New York Heart Association Class II. Electrocardiogram (ECG) and Holter monitor were normal but transthoracic echocardiography showed impaired left ventricular ejection fraction at 45%. Single-photon emission computed tomography (SPECT) myocardial perfusion imaging showed a perfusion

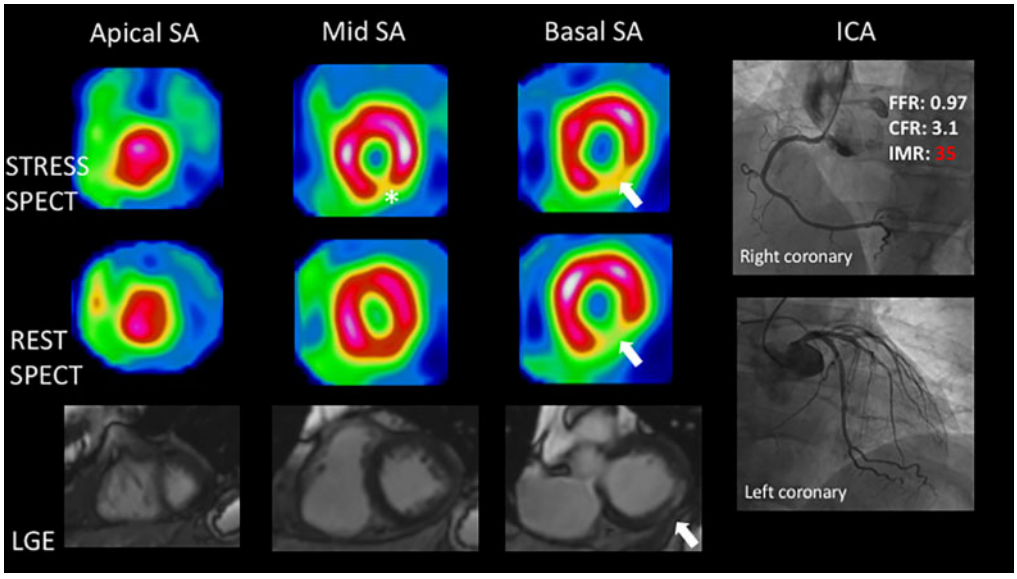

Figure 1 Single-photon emission computed tomography indicated a perfusion defect in mid and basal inferior segments with partial reversibility [mid-inferior defect at stress reversible at rest (white asterisk) and a basal inferior defect no reversible (white arrow)]. Invasive coronary angiography was negative for coronary stenosis. An abnormal index of microcirculatory resistance was observed in the ischaemic territory. Short-axis views of cardiac magnetic resonance images by late gadolinium enhancement imaging showed focal midwall myocardial fibrosis in basal inferior segment (white arrow). CFR, coronary flow reserve; FFR, fractional flow reserve; ICA, invasive coronary angiogram; IMR, index of microcirculatory resistance; LGE, late gadolinium enhancement; SPECT, single photon emission computed tomography; SA, Short Axis.

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defect in the mid and basal inferior segments with partial reversibility (*Figure 1*). The combination of ischaemia and ejection fraction impairment justified an ICA. Invasive coronary angiography indicated ischaemia in the absence of obstructive coronary artery disease, i.e. INOCA. The possibility of a false positive SPECT scan in the absence of actual ischaemia was investigated through the per-procedural assessment of the index of microcirculatory resistance (IMR) in the putatively ischaemic territory using an intracoronary pressure/temperature sensor-tipped guide wire (PressureWire X, Abbott, IL, USA). The results were as follows: fractional flow reserve: 0.97, coronary flow reserve: 3.1, IMR: 35 (*Figure 1*). The >25 IMR value indicated coronary microvascular dysfunction. Finally, to determine whether coronary microvascular dysfunction was due to DM1 or to the patient's cardiovascular risk factors, cardiac magnetic resonance imaging was performed and showed a focal midwall late gadolinium enhancement as typically observed in DM1 cardiomyopathy (*Figure 1*). In conclusion, the present case demonstrates the usefulness of IMR for the detection of coronary microvascular dysfunction in the setting of DM1 cardiomyopathy with INOCA.

Consent: The authors confirm that written consent for submission and publication of this case report including images and associated text has been obtained from the patient in line with COPE guidance.