Young women with acute coronary syndrome (ACS) frequently have nonatherosclerotic coronary artery disease (NACAD) and may be misdiagnosed. Coronary fibromuscular dysplasia (CFMD) commonly is overlooked, as the angiographic appearance is often subtle. Our group previously described CFMD as a diffuse obliterative disease starting abruptly at the mid-distal vessel, involving long segments.1 Spontaneous coronary dissection (SCAD) is another common form of NACAD and may be superimposed on CFMD, causing ACS. We report the first case series of SCAD and concomitant fibromuscular dysplasia (FMD).

Case 1 (Figure 1)
A 35-year-old African female with no cardiovascular (CV) risk factors presented on April 18, 2011 with persistent chest pain for 2 days while doing aerobic exercises. ECG showed T-inversion anteriorly and inferiorly, and troponin I (Tn-I) peaked at 4.2 μmol/L. Coronary angiogram showed diffuse severe stenosis from mid-right coronary artery (RCA) to the distal posterior descending artery with TIMI3 flow. A subsequent optical coherence tomography (OCT) showed coronary dissection with extensive intramural hematoma, and she was treated with stents. She had incidental bilateral external iliac artery FMD, and her carotid computer tomography angiography (CTA) showed a flap at the origin of the left carotid artery, prompting catheterization that confirmed FMD of both carotid arteries.

Case 2 (Figure 2)
A 48-year-old white female had chest pain on April 21, 2011 2 days after running 10 km. ECG showed nonspecific ST-T
changes, and Tn-I peaked at 2.8 μmol/L. She had no CV risk factors and was not pregnant. Angiography showed a well-demarcated mid-left anterior descending lesion, presumed to be atherosclerotic. This was stented, leaving a mild proximal stent edge stenosis, which was thought to be plaque shift. She had recurrent chest pain 9 days later, and repeat angiogram showed worse proximal stent edge stenosis. OCT showed intramural hematoma proximal to the stent and residual “tacked-up” hematoma in the wall of the stented segment, implicating SCAD, which caused her original ACS. She was treated with another stent. She had incidental diffuse external iliac FMD.

Case 3 (Figure 3)
A 45-year-old white female had chest pain a day after skiing on April 25, 2011. ECG showed T-inversion anteriorly, and Tn-I was 3.4 μmol/L. She had no CV risks and was not pregnant. Angiography showed 70% mid-left anterior descending stenosis, and OCT revealed SCAD with intramural hematoma. She underwent stenting. She had incidental mild external iliac FMD, and CTA showed mild FMD of the right vertebral artery at C1-2.

Case 4 (Figure 4)
A 48-year-old white female presented with chest pain at rest on July 6, 2011, with Tn-I 0.8 μmol/L and normal ECG. She had no CV risk factors. Angiography showed SCAD of a large obtuse marginal (OM) and the distal RCA, with abrupt transition from normal to diseased segments. She was treated conservatively. She has incidental mild right renal FMD, and head CTA showed FMD of bilateral carotid and vertebral arteries, with 2 mm pseudoaneurysm of the left internal carotid artery.

Case 5 (Figure 5)
A 48-year-old white female had chest pain after lifting her child on Sep 9, 2011. ECG showed diffuse T-inversion
anterolateral and inferiorly, and Tn-I peaked at 0.29. She had premature menopause, and a family history of coronary artery disease. Angiogram revealed distal left anterior descending dissection with TIMI3 flow and hypokinesis of the anterolateral wall. She also has diffuse narrowing of the mid-distal OM3 with corresponding localized akinesis of the inferior wall, suggesting healed dissection from prior infarction. She also has FMD of bilateral renal and external iliac FMD.

Case 6 (Figure 6)
A 42-year-old female developed chest pain 30 minutes after intercourse on Oct 22, 2011. Initial ECG showed anterior ST elevation, but it resolved spontaneously 20 minutes later. Tn-I peaked at 0.57. Angiogram showed mid-left anterior descending 80% stenosis with TIMI3 flow, and intramural hematoma was confirmed on OCT. She was treated conservatively given her chest pain and ECG resolution. She had incidental bilateral mild renal FMD, and CTA showed mild right cervical internal carotid and vertebral arteries FMD.

These women with SCAD had concomitant FMD of other territories, raising the suspicion that underlying CFMD predisposed them to dissection, precipitating ACS. 3 had angiographic appearance readily mistaken for atherosclerotic coronary artery disease (cases 2,3, and 6), thus a high index of suspicion and adjunctive imaging for NACAD is pertinent. This association of SCAD and FMD is not rare; in fact, we
reported that $\approx 30\%$ of young women with biomarker positive ACS have SCAD/CFMD. Although not all SCAD patients have CFMD, it is important to exclude FMD, as involvement of other vascular territories have long-term consequences; we routinely perform nonselective abdominal angiogram on digital subtraction (for renal and iliac) and head CTA. Empirically, they are treated with dual antiplatelet therapy and $\beta$-blockers per standard ACS guidelines. This novel association necessitates further studies to evaluate a potential causal link.

Figure 6. Case 6. A Coronary angiogram in postero-anterior cranial projection showing long mid-left anterior descending stenosis (*), which was caused by extensive intramural hematoma compressing the vessel lumen from SCAD (white arrows correspond to OCT images from C1–6). B Coronary angiogram in left anterior oblique cranial projection showing the well-demarcated appearance of this diffuse mid-left anterior descending stenosis (*). C OCT images corresponding to the arrows in A: C1 showing normal proximal left anterior descending segment, C2 showing intramural hematoma (*), C3 showing more compression of vessel lumen by intramural hematoma (*), C4 showing intimal thickening beyond the segment of intramural hematoma presumed to be caused by FMD involvement of the vessel wall (*), C5 showing calcification (*) distal to the intramural hematoma, also presumed to be caused by fibromuscular dysplasia (FMD) involvement of the vessel wall, and C6 showing normal vessel wall distal to the intramural hematoma. D ECG showing anterior ST elevation on presentation. E Right renal artery angiogram showing mild FMD involvement with mild beading-like appearance of the vessel wall (*).

Disclosures

None.

References

Spontaneous Coronary Artery Dissection in Patients With Fibromuscular Dysplasia: A Case Series
Jacqueline Saw, Rohan Poulter, Anthony Fung, David Wood, Jaap Hamburger and Christopher E. Buller