Behçet syndrome (BS) is an uncommon systemic vasculitis, with limited understanding of the pathophysiology and treatment options are nonspecific. Associated morbidity is higher in men, specifically those affected by vascular, neuronal, or ocular involvement. Systemic veins and pulmonary arteries are predominantly involved, both of which carrying deoxygenated blood at low pressure, presenting as occlusive (thrombotic/stenotic) or aneurysmal lesions. Arteries are also affected by similar lesions, because of inflammation of the vasa-vasorum resulting in medial defragmentation of the vessel wall. Stenotic lesions are considered benign, whereas aneurysmal change can be progressive and rupture can be fatal. Stenotic lesions are thought to be better treated with stenting, as surgical repair can result in aneurysmal change at the site of vascular injury.1

A 32-year-old male, known to have BS with the presence of antiphospholipid antibody, multiple episodes of deep vein thrombosis and pulmonary emboli but no apparent cardiac risk factors presented with exertional angina and abnormal stress test. At the time of index percutaneous coronary intervention, the BS was in remission with colchicine alone. Coronary angiography demonstrated total occlusion of the proximal left anterior descending artery that was successfully treated with percutaneous coronary coronary intervention, performed through bilateral transradial vascular access. BioMatrix (Biosensors, Inc, Newport Beach, CA), a biolimus-A9–eluting stents, were deployed extending from ostial left anterior descending artery into a good caliber diagonal artery, as we failed to wire into distal left anterior descending artery that received collateral blood supply from right coronary artery. He was commenced on aspirin and clopidogrel in addition to his long-term warfarin therapy. BioMatrix stents were chosen as they have an absorbable polymer and a relatively short drug release time allowing earlier endothelialization. We planned to repeat coronary angiography at 6 months with optical coherence tomography to assess stent struts coverage to ascertain, whether a shorter period on triple therapy was feasible. Repeat coronary angiography demonstrated aneurysm formation, only in the proximal segment of the vessel did not demonstrate similar change (Figure 1). Optical coherence tomography confirmed remodeling of the vessel wall, unapposed and uncovered stent struts in the aneurysmal segment (Figure 2E and 2F), whereas struts were well apposed and covered at proximal and distal ends (Figure 2B–2D and 2G). Interestingly, some stent struts appear covered despite of not being well apposed to the vessel wall that has remodeled. This suggests that the process of remodeling occurred at a slower rate than the stent strut coverage, resulting in the mound of endothelium (Figure 2E).

Meanwhile, the patient also developed 2 separate swellings on left forearm, confirmed to be radial artery aneurysms. The one was successfully treated with ultrasound-guided compression, whereas the other failed to respond fully. It was felt that the BS was more active than suggested by the blood tests and clinical picture; he was commenced on mycophenolate mofetil along with colchicine. We aim to avoid any further invasive interventions, including coronary angiography2 as any vascular trauma can lead to aneurysmal change at the site of the injury,3 risk of which can be possibly lowered, with the use of anti–tumor necrosis factor-α therapy cover. A computed tomographic scan of the thorax, abdomen, and pelvis along with duplex studies of the lower limb arteries reveals no other aneurysms. The radial artery aneurysm size has slightly decreased 12 months later and the patient remains angina free to date.

This case demonstrates rare, but occlusive lesion of the coronary artery in patient with BS that changes to aneurysm formation after vascular injury in the form of ballooning and stenting. Vasculitic activity in BS may be more than appreciated, despite the lack of clinical signs/symptoms and a normal inflammatory marker levels.

Disclosures
None.

References


Figure 1. Coronary angiography, pre and post angioplasty. Angiographic appearance of the left anterior descending artery (LAD), before percutaneous coronary intervention (PCI; A and D), immediately after PCI (B and E), and 5 months after PCI (G and F). Arrows marks the length of the occluded segment of LAD.

Figure 2. Optical coherence tomography (OCT) demonstrating vessel remodeling. Five months after percutaneous coronary intervention, angiographic appearance of the left anterior descending artery (LAD; A), frames of OCT in the stented segment from distal to proximal LAD (B–G), and left radial artery aneurysm (H). *Aneurysmal changes in coronary and radial arteries.

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