Healing of Coronary Artery Dissection in Connective Tissue Disease: The Dichotomy between Pathologic and Physiologic

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Abstract
The natural history of iatrogenic coronary dissections in the setting of scleroderma is not well described. This microvascular autoimmune process is rarely associated with premature atherosclerosis, thus leaving few cases of percutaneous coronary interventions (PCI) and their complications in the literature. We describe a case of a type D dissection of the left anterior descending artery (LAD) during PCI. After stenting the culprit lesion, a mid/distal LAD dissection was managed medically after the patient was chest pain free and hemodynamically stable. Left ventricular systolic dysfunction with an ejection fraction of 30-35% was noted at the conclusion of the procedure. Second look angiography performed 4 weeks later revealed a healed persistent dissection with two functional lumens giving rise to diagonal and septal branches separately. Follow up echocardiography showed normalization of systolic function. Previous studies have described increased risk of abrupt vessel closure, procedural failure and repeat PCI in patients with type D dissections. The unique milieu of an underlying collagen vascular disease may affect the healing of intramural architecture, vessel recovery and functionality. We hypothesize that the vasculopathy and systemic autoimmune process may have played a role in physiologic, but not pathologic healing of the vessel wall.

Keywords — coronary dissection, scleroderma, collagen vascular disease

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I. INTRODUCTION
Scleroderma is an autoimmune systemic disease that affects multiple organs including the heart. Cardiovascular manifestations of scleroderma are usually due to microvascular involvement. Acute coronary syndrome due to premature atherosclerosis is extremely rare in this population [1]. Although coronary artery dissection during PCI is not uncommon, the incidence and outcome is unknown in patients with underlying vasculopathy.

We present a unique case of a patient with scleroderma who presented with severe epicardial coronary artery disease and an acute anterior wall myocardial infarction. Emergent percutaneous intervention with stenting restored flow to the LAD, but was complicated by a Type D dissection that was managed medically. Repeat catheterization a few weeks later revealed a persistent dissection with two functional lumens and restoration of left ventricular systolic function. Progressive healing of dissections in the setting of connective tissue disease is not well described in the literature.

II. OUR EXPERIENCE
A 59-year old female presented to the emergency department with chest pain of four hours duration. Her past medical history was significant for Sjogren’s syndrome and scleroderma.

Figure 1. Diagnostic angiography revealed subtotal mid LAD culprit vessel with TIMI I flow on initial angiography.
She was complaining of typical chest pain, radiating to both arms and associated with nausea and vomiting. No traditional risk factors for atherosclerotic heart disease were noted. Her blood pressure was 107/66 mmHg, heart rate 82 beats per minute, respiratory rate 16 breaths per minute and she was afebrile. The cardiac exam was unremarkable. A 12 lead electrocardiogram (ECG) performed in the emergency department revealed normal sinus rhythm with 1 mm ST segment elevation in the anterior precordial leads and 1 mm ST segment depression in the inferior limb leads. The patient was taken for an emergent cardiac catheterization. Angiography revealed the culprit to be a 99% stenosis in the mid left anterior descending coronary artery with moderate diffuse calcification and TIMI I flow (Figure 1).

She underwent percutaneous intervention of the mid LAD. The lesion was crossed with a wire without difficulty and dilated with 2.0x15mm balloon. However, the wire appeared to be in a subintimal plane distally, so it was repositioned with distal intravascular placement confirmed via contrast injection. However, subsequent angiography was again suggestive of extraluminal position of the wire in the mid LAD. A second wire was placed in the “second lumen” and into the distal vessel with confirmation of intravascular position. A “dual lumen” appearance persisted in the mid vessel. A 2.5x24 mm bare metal stent was deployed at the site of the original lesion. After stent deployment a persistent type D dissection was seen beyond the distal edge of the stent with two distinct lumens appreciated (Figure 2 and Supplementary Video 1).

The decision was made to adopt conservative management, as the patient was chest pain free, hemodynamically stable and had TIMI III flow. Post PCI echocardiogram revealed a normal left ventricular size with severe hypokinesis of the anterior wall, septum and apex. The systolic function was estimated to be 30-35% and there was no evidence of pericardial effusion. She had an uneventful hospital course and was successfully discharged home in stable condition. She continued to do well with no active cardiovascular symptoms. A second-look coronary angiogram performed one month later demonstrated a patent stent in the mid LAD and persistent type D dissection in the distal vessel with 2 separate lumens with TIMI III flow (Figure 3 and Supplementary Video 2 and 3).

It appeared that the vessel had two functional lumens separated by an unhealed dissection flap. Branch vessels also filled via both the true and false lumens providing adequate flow to the myocardium. A follow up echocardiogram revealed normalization of the ventricular size and systolic function with an ejection fraction of 55%.

Figure 2. Type D dissection distal to the mid LAD stent after initial intervention (see also Supplementary Video 1).

Figure 3. Second look angiography demonstrated patent stent with persistent Type D dissection with TIMI III flow. Two functional lumens are visualized with an unhealed dissection flap between them (see also Supplementary Video 2 and 3).

III. DISCUSSION

Coronary artery dissection is common during percutaneous coronary intervention. It is more common in females, the elderly and heavily calcified and tortuous vessels [2]. The incidence of iatrogenic dissection in patients with underlying vasculopathy especially in the setting of connective tissue disease is ill defined. The hallmark of scleroderma is diffuse inflammation and fibrosis affecting multiple organs. The characteristic vascular involvement is microvascular dysfunction [3]. In the heart, histopathological examination typically reveals disseminated plaques of fibrosis, contraction bands of necrosis and normal epicardial coronary arteries with intimal hypertrophy [4]. Myocardial Raynaud’s phenomenon has been described as the cause of angina and myocardial infarction in those patients [5]. Additionally, it has been well accepted that the mechanism for underlying reversible perfusion defects on myocardial perfusion imaging is small
vessel vasospasm that responds well to vasodilators [6]. Premature and accelerated atherosclerosis and macrovascular coronary artery involvement is considered rare in the setting of connective tissue diseases [7,1,8].

In this case, our patient had no classic risk factors for atherosclerosis except for her age. The severe coronary disease and presentation of acute anterior myocardial infarction is unique given the patient’s pertinent medical history. After the percutaneous intervention to the LAD a distal Type D dissection was evident. Outcomes after dissection depend largely on the severity of intimal disruption. Classification of coronary artery dissection is based on angiographic appearance and ranges from Type A to F, as determined by the National Heart, Lung and Blood Institute (NHLBI) (Table 1). Mild to moderate dissections include type A and B, which are usually benign and regress spontaneously during follow up [9]. Types C through F are considered more significant with higher rates of major short and long term complications [10]. The management of these dissections is controversial with some studies showing that stenting of type C dissections does not improve long term outcomes [11]. On the contrary, it has been reported that abrupt vessel closure is three times more likely with type D spiral dissections compared to type A dissections.

Additionally, they have been associated with a significant decrease in procedural success and higher rates for repeat PCI, bypass surgery and Q-wave myocardial infarctions [12]. Many of the prior studies looking at prognosis of coronary artery dissection excluded patients with severe dissection [11,13]. Our patient had a type D coronary dissection beyond a short stented segment that was treated conservatively. It did not result in abrupt closure of the vessel. Moreover, it failed to heal and had the appearance of two “functional” lumens that maintained normal flow. This flow helped restore left ventricular function to normal despite initially documented hypokinesis of the anterior wall and moderate overall LV impairment.

### Table 1: Angiographic classification of coronary artery dissection from the national heart, lung and blood institute.

<table>
<thead>
<tr>
<th>Type</th>
<th>Description</th>
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<tbody>
<tr>
<td>Type A</td>
<td>Minor radiolucent area within coronary artery with little or no persistence of contrast</td>
</tr>
<tr>
<td>Type B</td>
<td>Parallel tract or double lumen with little or no persistence of contrast</td>
</tr>
<tr>
<td>Type C</td>
<td>Contrast outside the lumen (extra-luminal cap) with persistence of contrast</td>
</tr>
<tr>
<td>Type D</td>
<td>Spiral (barber shop pole) luminal filling defects</td>
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<tr>
<td>Type E</td>
<td>New persistent filling defect within the coronary lumen</td>
</tr>
<tr>
<td>Type F</td>
<td>Total occlusion of the coronary lumen without distal antegrade flow</td>
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</table>

The healing process of dissections can be unpredictable and eventually cause spontaneous improvement or deterioration by luminal narrowing and thrombosis [14]. The effect on intramural healing or recovery in the setting of scleroderma of Sjogren’s syndrome has not been well studied. Evaluation with repeated angiography in our patient one month after the initial intervention showed two separate functioning lumens with TIMI III flow. Contrast in branch vessels indicated preservation of the true and false lumens. The initial anterior wall hypokinesia was resolved on echocardiography four months after the infarction. Thus, there was likely a component of stunned myocardium and restoration of adequate flow to the myocytes that allowed for recovery of function. It is interesting to note that another factor that might predict the healing process is the reference vessel size. A larger vessel diameter has a higher probability of healing [14]. This does not explain the unusual healing process in our case where the mid and distal LAD was a small 2.5mm vessel.

### IV. CONCLUSIONS

The natural history of coronary artery dissection in patients with underlying vasculopathy as in our case has not been well characterized in the literature. Our hypothesis is that the pathological process of scleroderma, as well as vessel tortuosity and calcification may have precipitated the coronary artery dissection and influenced its unique healing. This resulted in physiologic, but not pathologic healing of the vessel. This case evokes the possibilities of different healing processes after a complex iatrogenic coronary artery dissection in a patient with connective tissue disease that has not been previously reported in the literature.

### V. SUPPLEMENTARY MATERIAL

Supplementary material is available at: http://www.researchpub.org/journal/jcvd/jcvd.html

- **Video 1**: 1\(^{st}\) cath video showing post stent result of residual Type D dissection;
- **Video 2**: 2\(^{nd}\) look angiography showing residual dissection with two functional lumens giving rise to diagonal and septal branches;
- **Video 3**: 2\(^{nd}\) look angiography in left anterior oblique (LAO) projections showing 2 functional lumens with residual dissection.

### REFERENCES