Concomitant coronary artery disease in a patient with Loeys-Dietz Syndrome and multiple aortic aneurysms

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Introduction

Loeys-Dietz syndrome (LDS) is a recently described connective tissue disorder characterized by early and aggressive onset of aneurysm formation and dissection of the aorta.[1] The lesions in these patients present early, often in childhood, and at much smaller aortic diameters.[2] Mean age of survival for patients diagnosed with LDS is 37 years.[3] These patients also have a typical phenotype: including a bifid uvula, hypertelorism, cleft palate, and vascular tortuosity derived from genetic alterations in TGFβ.[1, 3] Specifically, management of thoracic vascular abnormalities drives the treatment of these patients. We describe a patient with LDS who presented with multiple aortic aneurysms, cerebral aneurysms and surprisingly, concomitant coronary artery disease.

Case

A 52-year-old man presented to the emergency room with lower back pain. He had strained his back a few months previously and the pain never resolved. He eventually underwent an MRI and was found to have an eight centimeter abdominal aortic aneurysm. He was referred for aortic consultation and after further questioning, was found to have a significant family history. His brother had previously undergone successful valve sparing aortic root replacement and was diagnosed with LDS. Our patient had refused both genetic testing and imaging at the time of his brother's diagnosis. After genetic testing, he was found to have the TGFBR1 mutation, which was the same mutation as his brother, and the diagnosis of LDS was made. He underwent further imaging studies and was found to have a 5 cm aortic root aneurysm, 8 cm infra-renal aortic aneurysm, 7mm left cavernous internal carotid artery aneurysm and a 7 mm basilar artery aneurysm [Figure 1]. Because of size and symptoms, plans were made for initial abdominal aortic aneurysm repair, followed by subsequent aortic root replacement. He underwent successful open aorto-bi-iliac infra-renal aneurysm repair.

In preparation for his aortic root surgery, a cardiac catheterization was performed and a 75% stenosis of his proximal left anterior descending artery was found [Figure 2]. Echocardiogram confirmed the dilated aortic root and revealed a tricuspid aortic valve with mild aortic insufficiency.

After recovering from his abdominal aortic aneurysm repair, he underwent a valve sparing aortic root replacement with coronary artery bypass grafting. The aortic root was replaced with a Valsalva graft, the aortic valve spared and re-implanted in the fashion of a David IV procedure. The distal aortic (proximal to the innominate artery) and coronary button anastomoses were
patients with dissections of the coronary arteries,[4, 5] but never with a stenotic lesion requiring intervention. The prevalence of coronary artery disease is unknown in patients with LDS, likely because most patients with this syndrome do not progress to an age when coronary angiography would be indicated. When evaluating surgical vascular disease in these complex connective tissue patients, concomitant coronary disease cannot be dismissed, and should be investigated in appropriate patients based on established clinical guidelines.

**Conclusion**

We present a patient with LDS who had concomitant coronary artery disease, highlighting the value of a thorough workup in patients with complicated connective tissue disorders. This emphasizes the need to consider routine cardiac diagnoses and appropriate workup for patients with such vascular diseases.

**Disclaimers**

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**References**