CASE REPORT

A rare cause of recurrent aortic dissection

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We report the case of a 19-year-old man with a history of Loeys–Dietz syndrome (LDS), which was diagnosed when he had a Stanford type A aortic dissection. He also had multiple aneurysms including ones in the innominate, right common carotid, and right internal mammary arteries. He had had multiple procedures including Bentall's procedure, repeat sternotomy with complete arch and valve replacement, and coil embolization of internal mammary artery aneurysm in the past. His LDS was characterized by gene mutation for transforming growth factor-β receptor 1. He presented to our facility with sudden onset of back pain, radiating to the right shoulder and chest. He was diagnosed with Stanford type B aortic dissection and underwent thoracic aorta endovascular repair for his aortic dissection. This case represents the broad spectrum of pathology associated with LDS where even with regular surveillance and aggressive medical management the patient developed Stanford B aortic dissection.

Keywords: Aortic aneurysm, Aortic dissection, Aortic root replacement, Loeys–Dietz syndrome

Introduction

Loeys–Dietz Syndrome is an inherited connective tissue disorder of the vascular system. With no specific diagnostic criteria to recognize the syndrome, genetic testing is mandatory to identify transforming growth factor mutations. Most patients present with aortic aneurysmal rupture and/or dissection. Frequent monitoring with imaging is required to monitor such patients.

Case report

The patient was a 19-year-old man who in October 2013 had a Stanford type A aortic dissection with contained rupture and aortic root aneurysm (Fig. 1). He had Bentall's procedure with No. 25 St. Jude valved conduit and ascending aortic replacement with No. 24 Vaskutek branched graft. Genetic testing had revealed involvement of transforming growth factor β receptor 1 (TGFBR1) gene consistent with LDS. Imaging studies including computed tomography angiogram (CTA) and magnetic resonance angiogram of the head and

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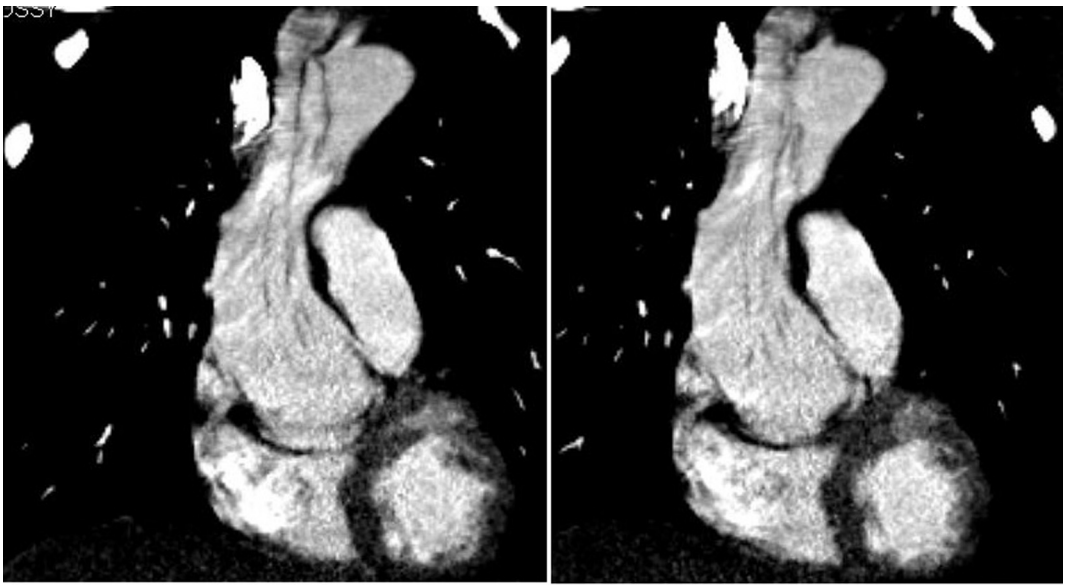

Figure 1. Computed tomography of chest, abdomen, and pelvis revealing Stanford type A aortic dissection with contained rupture and aortic root dissection extending into the right common carotid artery.

Figure 2. Computed tomography angiogram of the neck, chest and abdomen showing right common carotid artery and innominate artery aneurysm.

neck revealed multiple tortuous intracranial and cervical arteries consistent with LDS. He also had a 13 mm irregular right common carotid aneurysm.

In November 2013, when the patient had chest pain, concerns for new dissection arose and a

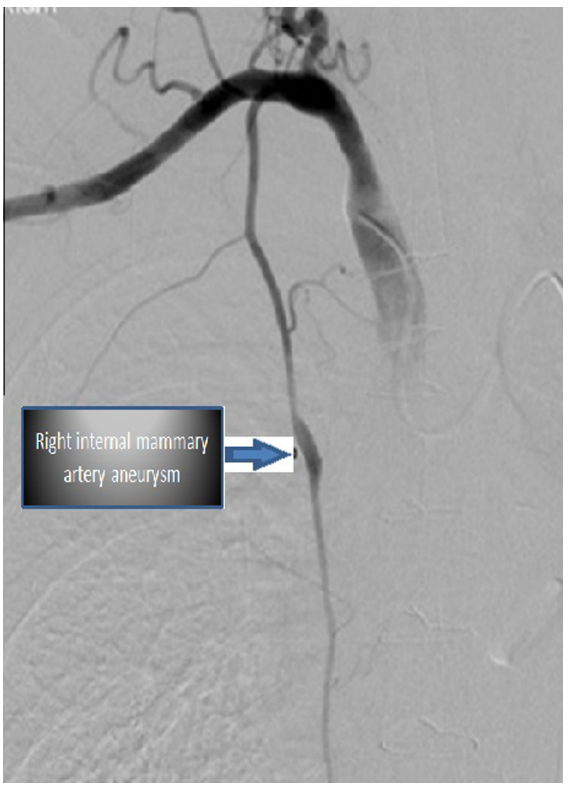

Figure 3. Fluoroscopy imaging showing right internal mammary artery aneurysm.