A case of Marfan's syndrome with multi-level aortic dissections

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Case Report

Abstract

BACKGROUND: Although Marfan's syndrome is a disease with various phenotypes, but the major mechanism of death is cardiovascular complication. Aortic dissection is a major cause of death in Marfan syndrome.

CASE REPORT: A 30-year-old man with severe refractory chest and left flank pain and history of previously surgically repaired Type A aortic dissection was referred to the hospital. His typical manifestations of Marfan's syndrome were identified. Cardiovascular imaging showed an acute spiral dissection in the descending aorta extending to the left renal and femoral arteries with no evidence of thrombosis in its huge false lumen (8 cm). By the diagnosis of acute, expanded, spiral, Type B aortic dissection, he underwent the stent grafting of dissected aorta. He discharged without any complication. On follow-up cardiovascular imaging, thrombosed false lumen in stented aorta from descending aorta to the proximal abdominal aorta was seen.

CONCLUSION: Endovascular treatment of Type B dissection is an effective treatment in Type B dissection, even in patients with Marfan syndrome.

Keywords: Marfan's Syndrome, Aortic Dissection, Endovascular Graft

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Introduction

The leading cause of morbidity and mortality in Marfan's syndrome is cardiovascular complication such as aortic root aneurysm and its subsequent rupture and dissection.[1,2] Here, we report a case with extensive Type B aortic dissection in a known case of Marfan's syndrome with history of surgical treatment of Type A aortic dissection.

Case Report

A 30-year-old male was referred to Emergency Department of Alzahra Teaching Hospital affiliated to Isfahan University of medical Sciences, Iran, with the chief complaint of persistent crescendo-decrescendo chest pain as well as severe refractory pain in the left flank. He has been a known case of Marfan's syndrome since 16 years ago, and it was initially diagnosed by bilateral lens dislocation, chest wall deformity (pectus excavatum), joint elasticity and vascular problems. The patient had previously undergone surgical repair for the diagnosis of Type A aortic dissection. On admission, the patient's hemodynamic status was stable and his vital signs were as follow: blood pressure of right and left arm (140-90 and 150/60 mmHg, respectively), heart rate: 78 bpm, respiratory rate: 30 and temperature: 37 °C. On physical examination, peripheral pulses were palpable. Pulses of left femoral artery, dorsalis pedis and posterior tibialis were weaker than those on the right. His laboratory data were as follow: blood urea nitrogen: 15.4 mg/dl, creatinine: 1.1 mg/dl, blood sugar: 145 mg/dl, white blood cells: 11,300/μ, platelet: 126,000/μ and hemoglobin: 16.6 mg/dl. Electrocardiography (ECG) showed incomplete right bundle branch block. Trans-thoracic echocardiography indicated moderate mitral regurgitation, moderate left atrial enlargement, moderate aortic insufficiency and

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dilated aortic root (4.45 cm) with previous dacron patch placement other ECG measures were within normal limit.

Patient was admitted with a diagnosis of aortic dissection. 64-slice computed tomography angiography (CTA) of thoracic and abdominal aorta was performed after intravenous injection of 100 ml of contrast agent. CTA of thoracic and abdominal aorta demonstrated evidences of flap and dissection originated adjacent the origin of left subclavian artery, extending to the proximal section of left femoral and left renal arteries without the presence of thrombosis in true/false lumens. A huge false lumen (8 cm) was seen. Superior mesenteric artery (SMA) was normally opacified, and celiac artery was originated from SMA as a normal variant. Right renal artery was well opacified and without any evidence of dilation (Figure 1).

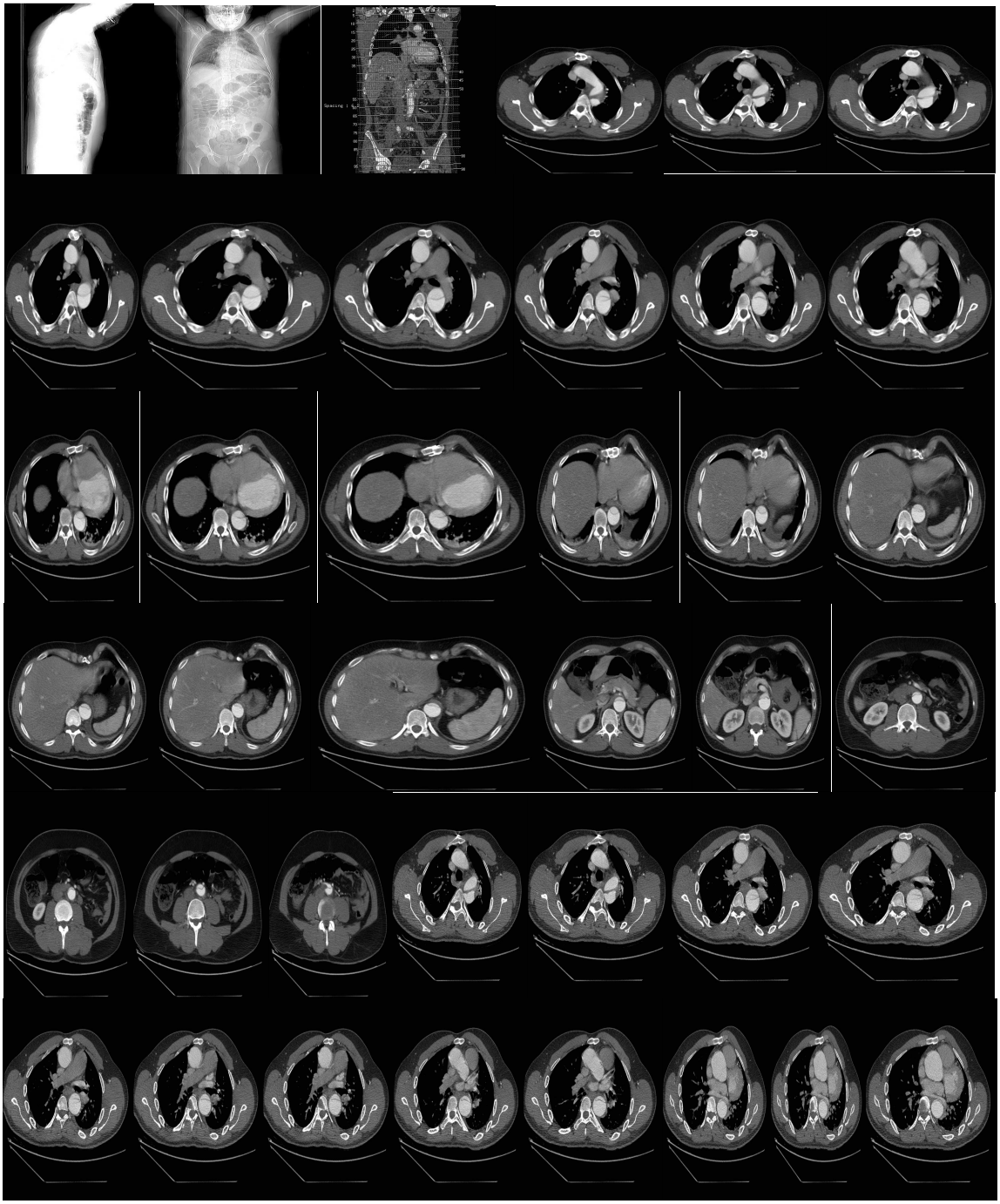

Figure 1. Computed tomography angiography before stent implantation

Aortic angiography showed spiral dissection originating from left subclavian artery and extended to iliac arteries (Video 1). Thus, the diagnosis of acute Type B dissection was confirmed.

He underwent endovascular repair by diagnosis of expanding Type B dissection. Through brachial and femoral access, the wire passed from true lumen and sized with a pig tail catheter was performed. Then, covered stent was previously deployed in ascending aorta. Subsequently, bare metal stent was deployed in descending aorta. Re-angiography of arch, ascending and abdominal arteries showed good results. In final injection, true lumen was seen, and both renal arteries were patented (Video 2). We decided to continue medical treatment for abdominal aorta dissection. After procedure, the force and movement of upper and lower extremities were normal. He was stable during hospitalization. In follow-up CTA, thrombotic false lumen was noted in descending aorta, associated with implanted stent which was placed from descending aortic arch down to the proximal part of abdominal aorta (just before SMA takes off). There was no evidence of aneurismal dilation in the thoracic aorta and major vessels originating from aorta (Figure 2). Left common iliac artery was involved in abdominal aorta dissection with good flow. The patient discharged on beta-blocker, angiotensin II receptor antagonist, aspirin, plavix and atorvastatin. He was serially followed-up after discharge. So far, he is asymptomatic.

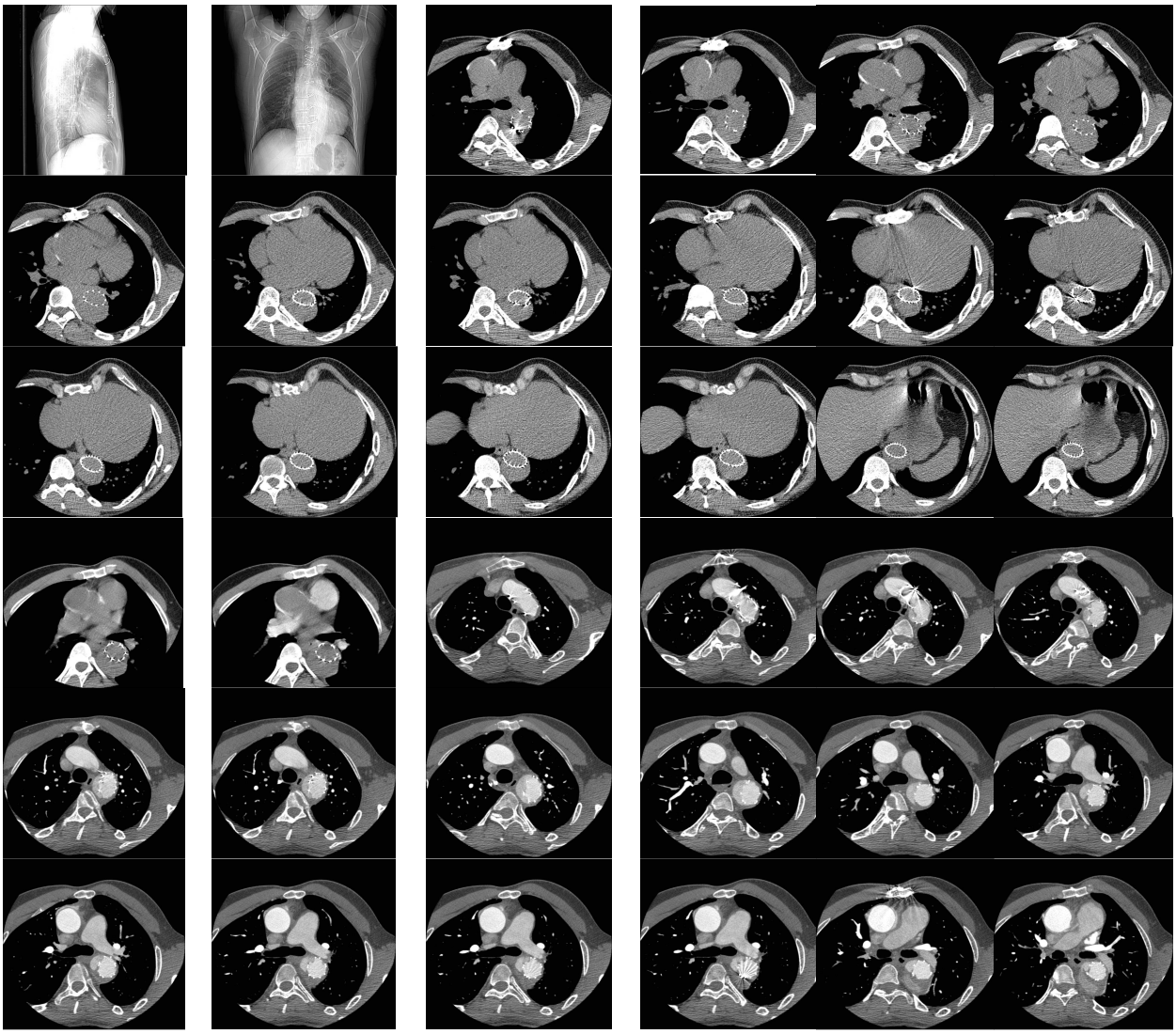

Figure 2. Computed tomography angiography after stent implantation

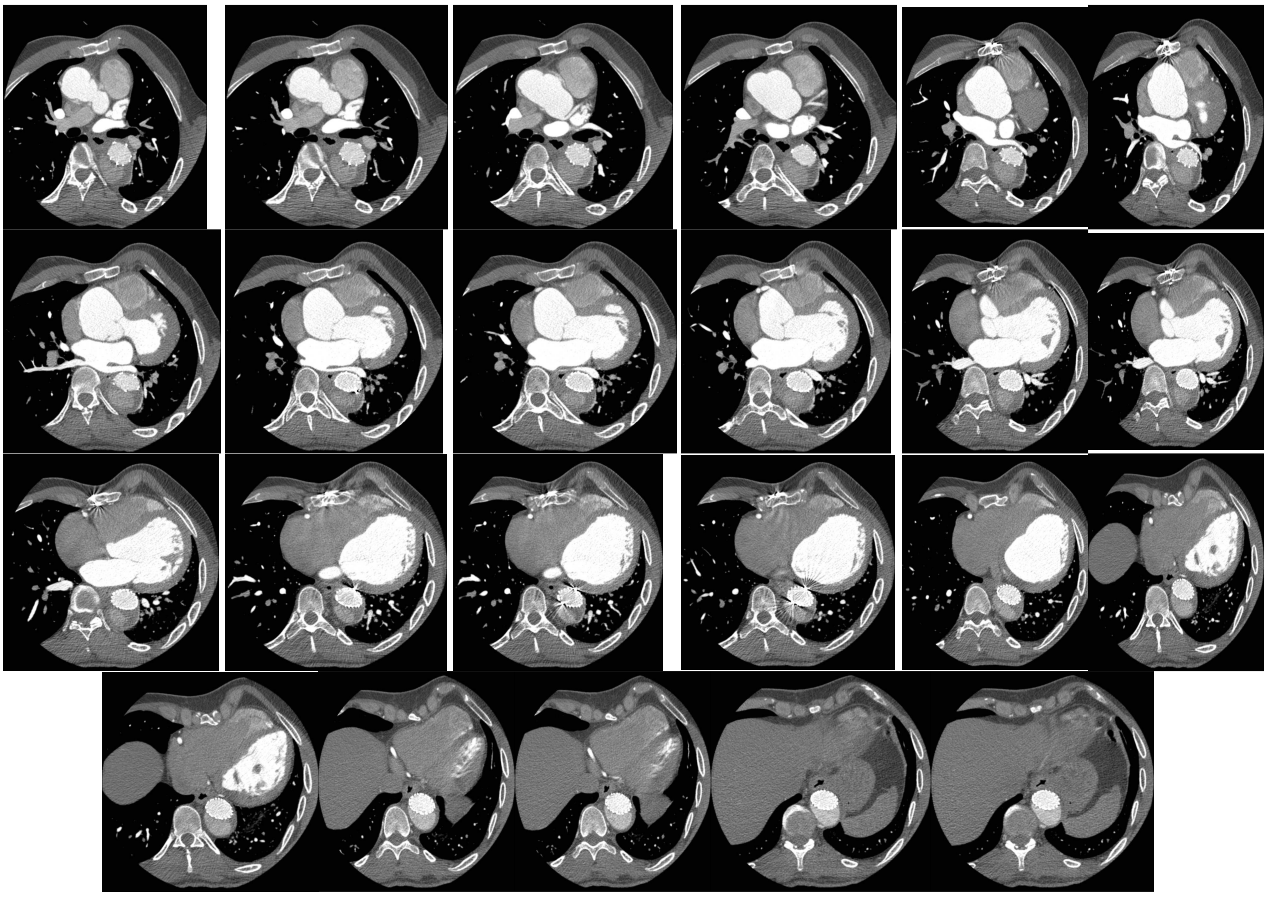

Figure 2. Computed tomography angiography after stent implantation (Continue)

Discussion

Marfan's syndrome, the most frequent inherited connective tissue disorder, is mainly associated with aortopathy such as aortic root dilatation or dissection and aortic valve insufficiency. Type B dissection is more prevalent in such patients' population. These cardiovascular complications limit life expectancy of the patients with Marfan's syndrome. By regular aortic follow-up and prophylactic aortic surgery, these catastrophic events are partly preventable.[3] Endovascular stent grafting in non-Marfan's syndrome patients with type B dissection and descending thoracic aorta is an established therapeutic procedure.[4] However in Marfan's syndrome patients, this procedure carries a significant propensity of endoleaks, surgical conversions and death.[3] Usually, endovascular treatments confront with early and late complications and these endovascular therapies need to be repeated. In this case, we performed a successful endovascular treatment and despite the huge false lumen, no need to repeated procedure happened and the patient was asymptomatic during the observation period. This misshapes mainly occur in Marfan's syndrome patients with chronic dissection.[5] Therefore, endovascular grafting should not be used routinely in such patients. The most of the Marfan's syndrome cases, aortic dissection is associated with aortic dilation. This patient experienced type A aortic dissection which was repaired by surgical treatment and this while he experienced new onset Type B dissection in the aortic location far from the previous site. Indeed, a huge non-thrombotic false lumen was seen in dissected portion that was successfully sealed by stent grafting.

Conclusion

Endovascular treatment of Type B aortic dissection is an elective treatment in type B dissection even in patients with Marfan's syndrome. Close future follow-up is recommended for these patients.

Acknowledgments

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Conflict of Interests

Authors have no conflict of interests.

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