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Asymptomatic, Chronic Type-A Dissection of a Large Ascending Thoracic Aortic Aneurysm in a Young Patient

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INTRODUCTION

Incidence of thoracic aortic dissection (TAD) in the general population is very low, ranging from 2.6 - 3.5 cases per 100,000 persons per year, but it is associated with a high rate of mortality and morbidity.[1-3] Based on the nature of its onset and anatomical location, TAD is classified as either acute or chronic Stanford type-A dissection involving the ascending aorta and type-B distal to the left subclavian artery.[4,5] Acute type-A dissection is highly lethal with a 30-day mortality of 50% compared to 10% of type B.[2] Most acute TAD patients presented with a sudden onset of severe chest, abdominal, or back pain, but 6.4% of them may have painless dissection.[6] The majority of patients with TAD were older with a mean age of 63 years while only 7% of them were less than 40 years of age.[7] Common predisposing factors for TAD are hypertension, atherosclerosis, and a history of cardiac surgery[2], while in young patients they are more likely Marfan's syndrome, bicuspid aortic valve, and prior aortic surgery.[7]

We report a case of a healthy, young male veteran who presented with asymptomatic, chronic type-A dissection of a large aortic aneurysm, complicated by severe aortic regurgitation (AR). Several physical signs characteristic of chronic, severe AR were found in this patient.

CASE REPORT

A 29-year-old Caucasian male with history of "heart murmur" diagnosed years prior was evaluated in the cardiology clinic as a referral from his primary care physician. Clinically, he was free of any symptoms even with routine exercise and work-out. He was a former smoker without history of recreational drug use. His past medical history included anxiety and migraine and a family history of "bicuspid aortic valve" with surgery in his paternal uncle (details unclear).

Physical examination showed that the patient was normal in appearance. His weight was 61.6 kg and a height of 69 inches giving a body mass index of 20.02 kg/m^2. His vital signs showed a heart rate of 79 beats/minute and blood pressure (BP) of 108/41 mmHg, indicating a widened pulse pressure of 67 mmHg (normal range: 30 - 50 mmHg). His radial and femoral artery pulses were present and equal on both sides. Cardiac examination showed a laterally displaced apical pulse by palpation and a loud, harsh systolic murmur and a 3/6 diastolic rumbling murmur in the precordium by auscultation. Other physical findings of his chronic AR with their respective eponyms were as follows:

- Palpation of his radial artery pulse showed rapidly swelling/falling pulse, further accentuated by wrist elevation above his head (Corrigan's pulse).
- In supine position, his brachial BP was measured at 117/53 mmHg compared to his popliteal one at 161/61 mmHg (Hill's sign).
- There was an exaggerated drop of brachial diastolic BP from 49 mmHg (BP: 109/49) in sitting and normal arm position to 36 mmHg (BP: 97/36) when the arm was raised to shoulder height (Mayne's sign).
- Visual examination of his thumb nail showed exaggerated nail bed pulsation or blanching when tip of nail bed was gently pressured (Quincke's sign).
- Auscultation over his femoral artery revealed a to-and-fro murmur generated by gentle compression of the stethoscope over the artery (Duroziez's sign).

Transthoracic echocardiogram (TTE) showed a moderately dilated left ventricle (LV) with LV ejection fraction of 50% and tricuspid aortic valve with severe AR by color-flow Doppler (Figure 1). Both his aortic root and sinuses of Valsalva were severely dilated to 5.6 and 6.8 cm respectively with intimal flaps from aortic dissection in the aortic root (Figures 2 and 3).

Chest computed tomography (CT) with contrast revealed severely dilated aortic root and proximal ascending aorta with maximal diameter of 6.4 cm extending for 8 cm before tapering quickly to normal dimension in the distal ascending aorta (Figure 4). Within the large aortic aneurysm, there were intimal flaps consistent with aortic dissection (Figures 5 and 6). The patient was referred to an academic medical center for further management. He was evaluated initially as an outpatient at that facility by a cardiothoracic surgeon and subsequently underwent successful aortic valve replacement using a St. Jude mechanical aortic valve and aortic aneurysm repair with aortic root replacement without complication. Intraoperatively, extensive dissection was identified in the ascending aorta above the valve, extending into the commissures of aortic valve AR. Pathology report showed "myxoid degeneration and fibrosis" of the resected aortic valve leaflets and aortic aneurysm.

DISCUSSION

Aortic dissection is defined as a splitting process that occurs in the aortic media.[2,4] The first 14-day period after onset has been designated the acute phase, because morbidity and mortality rates are the highest and surviving patients typically stabilize during that period. The pathophysiology of the aortic dissection includes aortic intimal tear or intramural hematoma. The most common predisposing factor in International Registry of Acute Aortic Dissection (IRAD) was hypertension (72%), followed by atherosclerosis (31%) and a history of cardiac surgery (18%).[2] Any mechanisms that weaken the medial layers of the aorta will lead to dissection that includes hypertension, connective tissue disorders (i.e., Marfan's syndrome, Ehlers-Danlos syndrome, bicuspid aortic valve), vasculitis (i.e., giant cell arteritis, Takayasu arteritis, Behcet's disease), chest-wall injury/motor vehicle accident, or iatrogenic factors like catheter intervention, prior cardiac surgery among other uncommon causes.

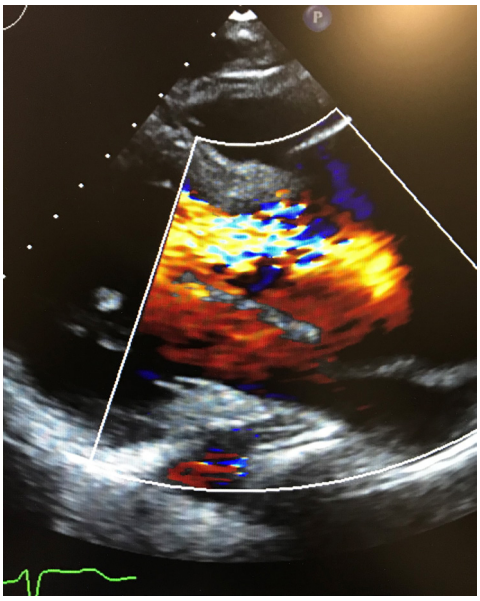

Figure 1. Transthoracic echocardiogram image in parasternal view showing severe aortic regurgitation by color flow Doppler.

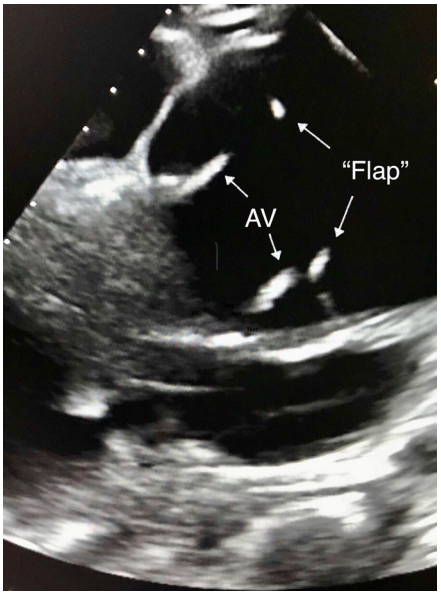

Figure 2. Transthoracic echocardiogram image in parasternal view of aortic valve in opening position and intimal flap in supravalvular position (both indicated by white arrowheads).

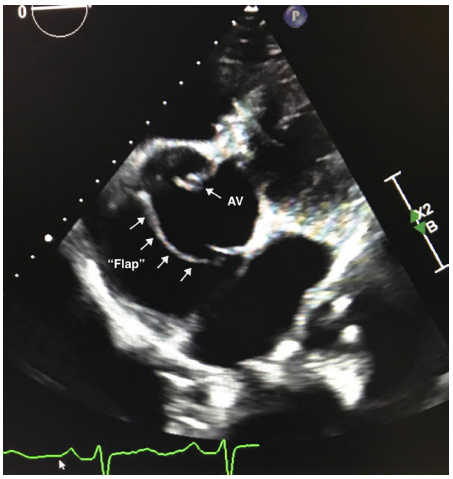

Figure 3. Transthoracic echocardiogram image in 5-chamber view showing severely dilated aortic root with intimal flap mimicking congenital supravalvular membrane.

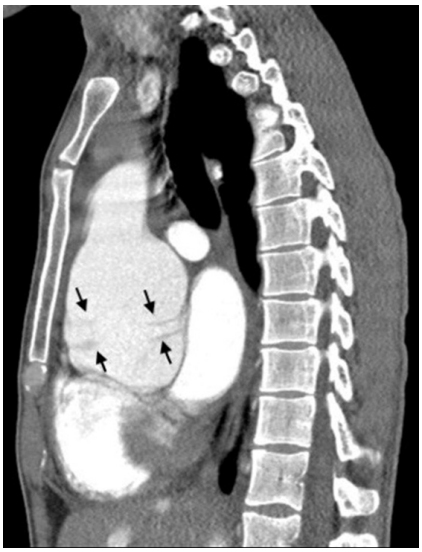

Figure 4. Sagittal view of chest computed tomography angiography showing a large proximal ascending aortic aneurysm, tapering down to normal dimension in distal ascending aorta. Arrowheads indicate intimal flaps.

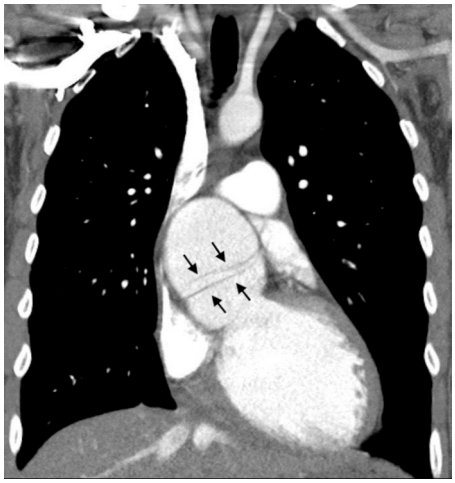

Figure 5. Coronal view of chest computed tomography angiography showing a large proximal, ascending aortic aneurysm. Arrowheads points to intimal flaps mimicking congenital supravalvular members.

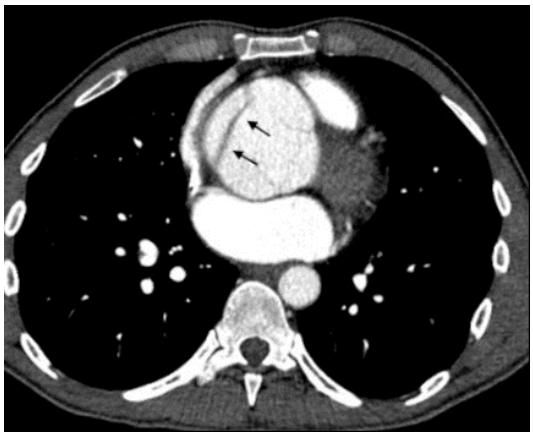

Figure 6. Axial view of chest computed tomography angiography showing dilated ascending aorta. Arrowheads indicate intimal flap.

Patients with TAD typically presented with sudden, severe chest, back, or abdominal pain characterized as sharp, tearing, or knifelike.[2,3] The chest pain was reported more common in type-A vs type-B dissection (78.9% vs. 62.9%) while abdominal pain was more common in type-B dissection (42.7% vs. 21.6% in type-A dissection).[3] Most TADs occurred in patients between 40-70 years of age, but no age group was exempt. However, it occurred much less frequently (less than 7%) in patients less than 40 years of age.[7] Overall in-hospital mortality was high (27.4%) in patients with acute thoracic dissection and highest among patients with type-A dissection (58.0%) not receiving surgery; patients with type-B dissection treated medically had the lowest mortality (10.7%).[2,3] In the IRAD study, the main reasons cited for medical therapy were comorbid conditions, advanced age (mean 80 years), and patient's refusal. The most common causes of death among patients with type-A dissection were aortic rupture, acute severe AR, cardiac tamponade, and visceral ischemia.[3,8] Upon follow-up of those patients who survived type-A dissection to discharge without surgical repair, 66% of patients had a 3-year survival rate.[8] Our patient likely had chronic TAD of undetermined duration. The pathology findings of myxoid, degenerative changes of his aortic valve leaflets and aorta were suggestive of underlying hereditary connective tissue disease.

Currently, the diagnostic modalities for TAD are transesophageal echocardiograph (TEE), CT, or magnetic resonance imaging (MRI) of the chest. For type-A TAD, abnormalities might be detected by TTE. The imaging hallmarks of dissection include the presence of an intimal flap dividing a true and a false lumen, intimal hematoma, pericardial effusion or tamponade, and aortic regurgitation.[9] Both TTE and chest CT findings of this patient were consistent with chronic type-A dissection of ascending thoracic aneurysm.

The one- and three-year survival for patients with acute type-A TAD treated with surgery was 96.1% and 90.5% vs. 88.6% and 68.7% without surgery.[8] Thus, surgery is the treatment of choice in patients with acute type-A TAD.

As a complication of TAD, extension of dissection to the aortic valve can result in AR. The hemodynamic consequences of AR are dependent on the rate of onset of AR, either acute or chronic.[10] In acute AR, there will be a sudden increase in the volume of blood in and increased filling pressure of the left ventricle (LV) leading to pulmonary edema and hypotension. Thus, severe, acute AR is a surgical emergency. If the individual survives the acute phase or has gradual worsening of AR, his LV adapts by hypertrophy and dilatation with a subsequent compensated volume overload and a normal LV filling pressure. Patients with chronic AR may be asymptomatic and may have normal exercise tolerance. In our patient, severe AR likely was due to extension of the dissection to the aortic valve commissure as noted intraoperatively.

Several physical findings were found in our patient characteristic of severe, chronic AR. These manifestations of severe chronic AR were the results of widened pulse pressure because elevated stroke volume exists during systole and the incompetent aortic valve allows the diastolic pressure within the aorta to fall significantly. A more comprehensive list[11,12] includes the following:

- Corrigan's pulse[13]: In 1832, Sir Dominic Corrigan (Dublin, Ireland) described a radial pulse, characterized by a rapidly swelling and falling by palpation, further accentuated by wrist elevation. It also was described later as "collapsing pulse" or "water-hammer pulse".
- Duroziez's sign[14]: Paul Duroziez, a French physician, in 1891 described the "double intermittent murmur" over the femoral arteries. It described a to-and-fro femoral artery murmur generated by femoral artery compression. This is likely caused by forward flow during systole and a diastolic flow from AR.
- Hill's sign[15]: Sir Leonard Hill, an English physiologist, first described the findings in 1909 that BP in the lower extremity in patients with AR was consistently higher than those in the upper extremities in recumbent position, without specifying numerical criteria. Later, a gradient above 20 mmHg generally has been accepted.
- Mayne's sign[16]: In 1953, Mayne described that a drop of at least 15 mmHg in the diastolic blood pressure when the arm was raised above the head could be detected in some of his patients with AR.
- Muller sign[11]: Systolic pulsations of the uvula are observed by visual examination.
- De Musset sign[17,18]: The sign of a bobbing motion of a patient's head with each heartbeat was named after a famous French writer and dentist, Alfred Louis Charles de Musset in 1877, who suffered from syphilitic aortitis and AR. It was described originally by his brother Paul[17] and later appeared in medical literature in 1900.[18]
- Quincke's sign[19]: It describes an exaggerated visible pulsation or blanching of red capillary color seen in nail beds when tip of the nail bed gently pressured. This was demonstrated recently by a video in a patient with severe AR.
- Taube sign ("pistol-shot" pulse): Booming systolic and diastolic sounds are auscultated over the femoral artery.

Keywords: thoracic aortic aneurysm, aortic dissection, aortic regurgitation, eponyms

CONCLUSIONS

Type-A aortic dissection, a highly lethal condition, can occur in apparently healthy, young patients with atypical presentation of epigastric/abdominal pain followed by asymptomatic clinical course despite the dissection and associated severe AR. Aortic regurgitation is a common complication of type-A dissection; careful physical examination may lead clinicians to the diagnosis at bedside.

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