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.1,4 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. 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. Common predisposing factors for TAD are hypertension, atherosclerosis, and a history of cardiac surgery, while in young patients they are more likely Marfan’s syndrome, bicuspid aortic valve, and prior aortic surgery.2

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/m². 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 (Quincke’s sign). 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%). 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.
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**: 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**: 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**: 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**: 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**: Systolic pulsations of the uvula are observed by visual examination.

- **De Musset sign**: 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 and later appeared in medical literature in 1900.\(^{18}\)

- **Quincke’s sign**: 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.
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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REFERENCES

1. Bickerstaff LK, Pairolero PC, Hollier LH, et al. Thoracic aortic aneurysms: A population-based study. Surgery 1982; 92(6):1103-1108. PMID: 7147188.
2. Tsai TT, Nienaber CA, Eagle KA. Acute aortic syndromes. Circulation 2005; 112(24):3802-3813. PMID: 16344407.
3. Hagan PG, Nienaber CA, Isselbacher EM, et al. The International Registry of Acute Aortic Dissection (IRAD): New insights into an old disease. JAMA 2000; 283(7):897-903. PMID: 10685714.
4. Crawford ES. The diagnosis and management of aortic dissection. JAMA 1990; 264(19):2537-2541. PMID: 2232021.
5. Daily PO, Trueblood HW, Stinson EF, Wuerflein RD, Shumway NE. Management of acute aortic dissections. Ann Thorac Surg 1970; 10(5):237-247. PMID: 5458238.
6. Park SW, Hutchison S, Mehta RH, et al. Association of painless acute aortic dissection with increased mortality. Mayo Clin Proc 2004; 79(10):1252-1275. PMID: 15473405.
7. Januzzi JL, Isselbacher EM, Fattori R, et al. Characterizing the young patient with aortic dissection: Results from the International Registry of Aortic Dissection (IRAD). J Am Coll Cardiol 2004; 43(4):665-669. PMID: 14975480.
8. Tsai TT, Evangelista A, Nienaber CA, et al. Long-term survival in patients presenting with type A acute aortic dissection: Insights from the International Registry of Acute Aortic Dissection (IRAD). Circulation 2006; 114(suppl 1):E350-E356. PMID: 16820599.
9. Flachskampf FA. Assessment of aortic dissection and hematoma. Semin Cardiothorac Vasc Anesth 2006; 10(1):83-88. PMID: 16703240.
10. Nishimura RA, Otto CM, Bonow RO, et al. 2014 AHA/ACC guideline for management of patients with valvular heart disease: A report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines. J Am Coll Cardiol 2014; 63(22):e57-e185. PMID: 24603191.
11. Ashrafian H. Pulsatile pseudo-protosis, aortic regurgitation and 31 eponyms. Int J Cardiol 2006; 107(3):421-423. PMID: 16503268.
12. Babu AN, Kynes SM, Carpenter Fryer SM. Eponyms and the diagnosis of aortic regurgitation: What says the evidence? Ann Intern Med 2003; 138(9):736-742. PMID: 12729428.
13. Corrigan DJ. On permanent patency of the mouth of the aorta, or inadequacy of the aortic valves. Edinb Med Surg J 1832; 37(111):225-245. PMID: 30331470.
14. Major RH. Classic descriptions of disease. 3rd Ed. Springfield, II: Charles C Thomas, 1945, p. 364. ISBN: 0-398-01202-4.
15. Hill L, Flack M, Holtzmann W. The measurement of systolic blood pressure in man. Heart 1909; 1:73-82.
16. Mayne B. On aortic regurgitation: A new physical sign. Ir J Med Sci 1953; (326):80-81. PMID: 13034347.
17. Sapira JD. Quincke, de Musset, Duroziez, and Hill: Some aortic regurgitations. South Med J 1981; 74(4):459-467. PMID: 7013091.
18. Delpeuch A. Le signe de Musset: secousses rythmiques de la tete chez les aortiques (de Musset’s sign: a rhythmic bobbing of the head in aortic cases). Presse Med 1900; 8:237-238.
19. Vindhyal MR, Vindhyal S, Boppana VS, Quincke’s pulse. Kans J Med 2019; 12(2):55. PMID: 3119812.