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MINI-FOCUS ISSUE: VASCULAR MEDICINE

BEGINNER

CASE REPORT: CLINICAL CASE

Pseudoaneurysm as a Late Complication of Chronic Stanford Type A Intramural Hematoma Requiring Endovascular Repair

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ABSTRACT

Aortic intramural hematoma accounts for 5% to 20% of patients with acute aortic syndrome. Endovascular grafts have evolved as minimally invasive alternatives for treatment in some highly selected patients. We present the case of a patient who had late complications of a chronic Stanford type A intramural hematoma requiring thoracic endovascular aortic repair. (Level of Difficulty: Beginner.) (J Am Coll Cardiol Case Rep 2020;2:2470-5) © 2020 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

HISTORY OF PRESENT ILLNESS

An 83-year-old man presented with acute onset abdominal pain that subsequently radiated to his chest and back. Symptoms persisted for nearly 12 h, at

LEARNING OBJECTIVES

- To appreciate potential late complications of Stanford type A IMH.
- To better understand the utility of TEVAR in treating patients with pseudoaneurysm formation as a late complication of Stanford type A IMH when performed at high-volume centers by multidisciplinary teams with experience in these procedures.
- To understand the importance of strict follow-up of patients with acute aortic syndrome, especially when managed conservatively.

which point he decided to come to the emergency department for evaluation. On arrival, his blood pressure was 219/126 mm Hg and was equal in both arms. An electrocardiogram showed sinus rhythm with first-degree atrioventricular block, nonspecific T-wave changes, and a heart rate of 86 beats/min. Results of the first set of cardiac enzyme tests were negative. The chest radiograph showed no widening of the mediastinum. He received intravenous labetalol and morphine, with only a modest improvement in symptoms.

Computed tomography (CT) angiography of the chest, abdomen, and pelvis was planned. The estimated glomerular filtration rate on admission was 18 ml/min. We did not proceed with the study given the patient's concerns about possible contrastinduced nephropathy and subsequent renal failure. An emergency transesophageal echocardiogram was performed. The images showed an intramural

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hematoma (IMH) extending from the ascending aorta through the aortic arch and propagating down the descending thoracic aorta at least to the origin of the celiac artery. Left ventricular size and systolic function were normal. There was a small circumferential pericardial effusion (**Figure 1**, Videos 1 and 2).

PAST MEDICAL HISTORY

Past medical history included resistant hypertension, traumatic subdural hematoma, coronary artery disease, prostate cancer, stage 4 chronic kidney disease, and hyperlipidemia.

DIFFERENTIAL DIAGNOSIS

The differential diagnosis included type A dissection, expanding thoracic aortic aneurysm, pseudoaneurysm, acute coronary syndrome, and pancreatitis.

INVESTIGATIONS

Following the transesophageal echocardiogram, noncontrast CT of the chest, abdomen, and pelvis was ordered. Images showed that the intramural hematoma originated at the aortic root and propagated to the descending thoracic aorta (Figure 2).

MANAGEMENT

The patient was admitted to the intensive care unit for treatment of Stanford Type A IMH. He was started on a labetalol infusion that was titrated to maintain a blood pressure of <120/80 mm Hg. Cardiothoracic surgery was consulted to evaluate the patient. The calculated risk of in-hospital mortality for a non-coronary artery bypass graft major procedure was 16.85% on the basis of the EuroSCORE II score (1). This was discussed with the patient, and a decision was made to follow a more conservative

approach. We were able to transition to oral labetalol, 600 mg 3 times a day, and amlodipine, 10 mg once daily. He was subsequently transferred to the telemetry floor where, on the sixth day of his hospital stay, he started to report dyspnea at rest. An audible wheeze was appreciated on physical examination. A subsequent chest radiograph showed clear lung fields, but it also revealed widening of the mediastinum that was not present on the initial study. A transthoracic echocardiogram was performed and showed no change in the luminal diameter of the ascending aorta, but a possible increase in thickness.

We considered possible intervention by thoracic endovascular aortic repair (TEVAR). He was thoroughly evaluated but again was deemed too high a risk for surgery. Blood pressure and renal function remained stable, and he was discharged home.

He was seen regularly in follow-up with office visits every 1 to 3 months. Serial imaging was not performed during these visits because the patient had expressed his wishes not to have any further aggressive measures taken. However, 1 year after his initial hospitalization, he presented with unexplained hypotension. A transthoracic echocardiogram was performed and showed that the ascending aortic



ABBREVIATIONS AND ACRONYMS

CT = computed tomography IMH = intramural hematoma TEVAR = thoracic endovascular aortic repair





FIGURE 3 Repeat Transthoracic Echocardiography Showing That the Ascending Aortic Aneurysm Is Now 6.7 cm in Diameter aneurysm had increased in size, with a new diameter of 6.7 cm and a possible dissection flap (Figure 3). He was sent on an emergency basis to the hospital for further care. Initial noncontrast CT showed aneurysmal dilation of the ascending aorta that was 7.46 cm in greatest diameter and a possible dissection flap (Figure 4). A subsequent magnetic resonance angiogram with contrast confirmed the presence of a Stanford type A dissection, but it also revealed a pseudoaneurysm of the ascending aorta (Figure 5, Video 3). He was admitted to the intensive care unit, and a labetalol infusion was initiated in preparation for transcatheter repair.

The aortic valve was crossed, and a Lunderquist wire (Cook Medical, Bloomington, Indiana) was placed in the left ventricle. The first endograft, Cook Alpha $42 \times 42 \times 80$ mm (proximal), was advanced to the ascending aorta over the stiff wire. The correct



position was verified, and the graft was deployed just above the sinotubular junction under rapid pacing, thus providing 2 cm of seal. A second graft, Cook Alpha $42 \times 42 \times 80$ mm (distal), was placed after removal of the delivery system of the first stent graft and was similarly deployed with considerable overlap, landing 1 cm proximal to the innominate artery. The overlap portion was ballooned with a 46-mm CODA balloon (Cook Medical) (Videos 4 and 5). Two stents were used, as opposed to a single longer stent, to prevent a "bird's beak" where the proximal portion of the stent graft can be very angulated. Using 2 stents also allowed for better conformation of the curvature of the ascending aorta and to prevent late graft failure. The Cook stent was chosen because of its low profile and stable deployment. It can also be modified and cut to shorter lengths if needed. Balloon expansion was not performed at the proximal or distal FIGURE 5 Magnetic Resonance Angiography of Stanford Type A Dissection With Pseudoaneurysm



FIGURE 6 Post-Operative Noncontrast Computed Tomography Images Showing Thoracic Endovascular Aortic Repair Present in the Ascending Aorta

F = foot; H = head.

segments of the stent graft to reduce the risk of creating a type A or B dissection.

A renal angiogram was performed that revealed severe ostial stenosis of the left renal artery. The decision was made to proceed with renal artery angioplasty given the patient's history of resistant hypertension that likely led to his acute aortic syndrome. Angioplasty was performed with a 4 mm \times 40 mm balloon. Residual stenosis was noted. The left renal artery was then stented with a pair of 6 mm \times 14 mm balloon expandable stents.

DISCUSSION

Aortic IMH accounts for 5% to 20% of patients with acute aortic syndrome (2). Aortic IMH is defined as a hematoma confined to the tunica media of the aorta in the absence of a detectable intimal tear (3). Some investigators have suggested that IMH represents acute aortic dissection where thrombus has formed to obliterate the false lumen (4). Fewer than 10% of these cases resolve spontaneously, with 16% to 47% progressing to dissection. Mussa et al. (5) evaluated 6 studies including 309 patients with either type A or type B IMH. The 30-day mortality rates were 4% to 19% for medical management, 11% to 24% for open surgical repair, and 0% to 6% for TEVAR.

In this case, the patient's age and comorbidities put him at a significantly increased risk of complications from open repair. In a comprehensive review performed by Muetterties et al. (6), the most common diseases of the ascending aorta treated with TEVAR were dissection, followed by pseudoaneurysm, penetrating aortic ulcer, aortic rupture, and aneurysm. TEVAR for the treatment of IMH is relatively less common (7,8). Our patient had an exceedingly rare aortic syndrome of ascending aortic aneurysm of 6.5 cm, dissection, intramural hematoma, and pseudoaneurysm. Also of interest was the timing of his intervention, almost 1 year after his initial presentation. He was successfully treated with TEVAR without any complications.

FOLLOW-UP

At the time this paper was written, 4 months after his operation, the patient remained hemodynamically stable. He tolerated the procedure well. We continue to manage his blood pressure with labetalol 300 mg three times daily, amlodipine 10 mg daily, and torsemide 10 mg daily. His post-operative noncontrast CT scan showed good endograft expansion and no pericardial effusion (Figure 6). We plan to perform a magnetic resonance angiogram of the chest in the future to evaluate for aortic graft endoleak.

CONCLUSIONS

When performed by an experienced multidisciplinary team, TEVAR may be a reasonable alternative to surgical repair for patients with serious ascending aortic disease and who have a high operative risk.

AUTHOR DISCLOSURES

The authors have reported that they have no relationships relevant to the contents of this paper to disclose.

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REFERENCES

1. Nashef SA, Roques F, Sharples LD, et al. Euro-SCORE II. Eur J Cardiothorac Surg 2012;41:734–45.

2. Maraj R, Rerkpattanapipat P, Jacobs LE, et al. Meta-analysis of 143 reported cases of aortic intramural hematoma. Am J Cardiol 2000;86: 664.

3. Al Rstum Z, Tanaka A, Eisenberg SB, Estrera AL. Optimal timing of type A intramural hematoma repair. Ann Cardiothorac Surg 2019;8: 524–30.

4. Uchida K, Imoto K, Karube N, et al. Intramural haematoma should be referred to as thrombosed-

type aortic dissection. Eur J Cardiothorac Surg 2013;44:366.

5. Mussa FF, Horton JD, Moridzadeh R, Nicholson J, Trimarchi S, Eagle KA. Acute aortic dissection and intramural hematoma: a systematic review. JAMA 2016;316:754-63.

6. Muetterties CE, Menon R, Wheatley GH 3rd. A systematic review of primary endovascular repair of the ascending aorta. J Vasc Surg 2018;67:332-42.

7. Chen YY, Yen HT, Wu CC, Huang DK. Thoracic endovascular aortic repair for type A intramural hematoma and retrograde thrombosed type A aortic dissection: a single-center experience. Ann Vasc Surg 2020;65:224–31.

8. Plichta RP, Hughes GC. Thoracic endovascular aortic repair for the ascending aorta: experience and pitfalls. J Vis Surg 2018;4:92.

KEY WORDS aorta, complication, dissection, vascular disease

PAPPENDIX For supplemental videos, please see the online version of this paper.