Primary aortic thrombus (PAT) in the absence of underlying aortic pathology such as atherosclerosis or aneurysm is quite rare and presents with various symptoms related to distal embolization. The symptoms can vary depending on the location of the PAT and organs affected. Although the definitive cause of thrombus formation is usually unknown, many patients have an underlying hypercoagulable state. Some physicians prefer conservative management because of their concerns about possible recurrence and questionable benefits from a complex operation; however, a delay or failure in conservative treatment can cause serious complications. Interventional treatment options include open surgical thrombectomy and thoracic endovascular aortic repair (TEVAR). The optimal management remains controversial, and there is no clear consensus.

METHODS

Between October 2016 and December 2020, a total of 10 patients were admitted to our institution with evidence of PAT. We reviewed medical charts, cardiovascular images, and operative records to identify patient characteristics, end-organ dysfunction, operative techniques used, and surgical results. Mid-term outcomes were determined from clinic records when available or from written correspondence with the patients’ physicians. This study was approved by the Temple University Institutional Review Board (protocol 28,911,
The baseline patient characteristics are shown in Table 1. Of the 10 patients, 6 were female, and the mean age was 49.1 ± 9.4 years (median, 48 years). Obesity was very common (mean body mass index [BMI], 32.1 ± 5.2; median, 33.6); in fact, 7 patients had a BMI >30. All the patients were active smokers at the time of initial presentation, and 6 patients had a hypercoagulable state, including Leiden V factor in 2, idiopathic thrombocytosis in 2, and hyperhomocysteinemia in 2. Two patients had antinuclear antibodies, and 1 patient had decreased levels of both antithrombin III and protein S. Two patients were active oral substance abusers. One patient was on highly active antiretroviral therapy for HIV infection. None of the female patients was on oral contraceptives.

PAT locations included the ascending aorta in 4 patients, the descending thoracic aorta in 3 patients, and the aortic root, aortic arch, and thoracoabdominal aorta in 1 patient each. Two of these patients had multiple thrombi. A distal embolization was common, especially in those who had PAT in the descending thoracic aorta (Table 1).

Surgical Approach and Intraoperative Findings

All open surgeries were performed under standard cardiopulmonary bypass. In terms of surgical approach, a standard median sternotomy was used for open thrombectomy of a PAT located in the aortic root and the ascending aorta, whereas a left thoracotomy approach was used for a PAT in the descending thoracic aorta. One patient (patient 3; Figure 1), who had multiple large thrombi on both the descending thoracic aorta and the abdominal aorta, underwent thoracoabdominal spiral incision. Intraoperative direct aortic ultrasound study (epiaortic echo) was often used for accurate identification of locations. Care was taken to avoid overmanipulating the aorta, to prevent dislodgement of the thrombus. Under aortic cross-clamping on the distal ascending aorta followed by cardiac arrest with cardioplegia in sternotomy cases, with aortic cross-clamping on both the proximal and distal sides of the aorta in thoracotomy cases with partial cardiopulmonary bypass, the aorta was opened and the thrombus gently removed. The intima of the aorta was carefully inspected to ensure that no residual component of the thrombus remained, and the inside of the aorta was copiously irrigated with saline solution. In some cases (patients 4 and 6), a small piece of the aortic wall was also resected along with the attachment of the thrombus, and this was repaired with either direct suture closure or interposition with a Dacron vascular graft. Also, when necessary, the aortotomy incision was closed using a Dacron patch to prevent any significant narrowing of the aorta (patients 1 and 3). No patient required deep hypothermic circulatory arrest.

One patient (patient 2; Figure 2), who presented with ST elevation myocardial infarction (MI), underwent emergency surgery. A coronary angiogram prior to surgery showed total occlusion of the right coronary artery due to embolization from a large thrombus located in the sinus of Valsalva of the aortic root. Another patient (patient 8; Figure 3) who presented with non–ST elevation MI had severe proximal left anterior descending coronary artery stenosis, apparently due to a thrombus, along with a large, highly mobile thrombus in the distal ascending aorta. These 2 patients underwent concomitant coronary artery bypass grafting in addition to urgent open thrombectomy.

One patient (patient 4; Figure 4), who presented with acute onset of hemiplegia and was diagnosed with cerebral infarction due to total occlusion of the right middle cerebral artery, underwent an emergency catheter-based mechanical thrombectomy by a neurosurgical team. CTA revealed a highly mobile large thrombus in the ascending aorta. Several days later, his ischemic stroke developed a hemorrhagic transformation. The decision was made to proceed with an open surgical thrombectomy of the PAT in the ascending aorta, given that his imaging and neurologic status remained stable for 1 week after development of the hemorrhagic transformation. Interestingly, this patient was found to have massive bilateral pulmonary emboli (PE) on intraoperative TEE and thus

<table>
<thead>
<tr>
<th>Abbreviations and Acronyms</th>
<th>Definition</th>
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</thead>
<tbody>
<tr>
<td>BMI</td>
<td>body mass index</td>
</tr>
<tr>
<td>CTA</td>
<td>Computed tomography angiography</td>
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<tr>
<td>HIV</td>
<td>human immunodeficiency virus</td>
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<td>PAT</td>
<td>primary aortic thrombus</td>
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<tr>
<td>PE</td>
<td>pulmonary embolism</td>
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<tr>
<td>SCA</td>
<td>subclavian artery</td>
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<tr>
<td>SMA</td>
<td>superior mesenteric artery</td>
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<tr>
<td>TEE</td>
<td>transesophageal echocardiography</td>
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approved November 2, 2021). The requirement for patient consent was waived.

Indications

At the time of clinical presentation, all patients underwent computed tomography angiography (CTA), as well as transthoracic and/or transesophageal echocardiography (TEE) for definitive diagnosis. The presence of an intracardiac thrombus was first ruled out. The exact locations and characteristics of the PAT were evaluated with these imaging studies. Surgical decisions were made depending on the location, size, and characteristics of the thrombus; end-organ dysfunction; and/or poor response/resistance to medical management, including intravenous (IV) anticoagulation. All patients underwent a routine hematology consultation to screen for a hypercoagulable state.

Patient Follow-up

Patients were followed up for 2 to 4 weeks by the attending surgeon at an outpatient clinic and every 3 to 6 months by the referring physician thereafter. Additional imaging studies were performed when any further symptoms developed.

RESULTS

Patient Characteristics

The baseline patient characteristics are shown in Table 1. Of the 10 patients, 6 were female, and the mean age was 49.1 ± 9.4 years (median, 48 years). Obesity was very common (mean body mass index [BMI], 32.1 ± 5.2; median, 33.6); in fact, 7 patients had a BMI >30. All the patients were active smokers at the time of initial presentation, and 6 patients had a hypercoagulable state, including Leiden V factor in 2, idiopathic thrombocytosis in 2, and hyperhomocysteinemia in 2. Two patients had antinuclear antibodies, and 1 patient had decreased levels of both antithrombin III and protein S. Two patients were active oral substance abusers. One patient was on highly active antiretroviral therapy for HIV infection. None of the female patients was on oral contraceptives.

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<table>
<thead>
<tr>
<th>Patient</th>
<th>Age, y/sex</th>
<th>Location and characteristics</th>
<th>End organ involved</th>
<th>Comorbidities /risk factors</th>
<th>BMI</th>
<th>Surgical procedure</th>
<th>Postoperative antiplatelets/anticoagulation</th>
<th>Late events</th>
<th>Follow-up, mo</th>
</tr>
</thead>
<tbody>
<tr>
<td>1 2016.10</td>
<td>56/F</td>
<td>Descending thoracic aorta × 3 (13, 14, 9 mm, mobile)</td>
<td>Spleen, kidney, LE</td>
<td>Factor V Leiden (heterozygous), anticardiolipin IgM, thrombocytosis (&gt;1000 K)</td>
<td>23.4</td>
<td>Left thoracotomy, thrombectomy with and without patch repair</td>
<td>ASA, rivaroxaban</td>
<td>Superficial wound dehiscence</td>
<td>2</td>
</tr>
<tr>
<td>2 2016.12</td>
<td>49/F</td>
<td>Aortic root (right coronary sinus, 23 mm, sessile)</td>
<td>STEMI, (RCA) TIA</td>
<td>HIV on HAART</td>
<td>38.4</td>
<td>Sternotomy, thrombectomy with direct aortic repair, CABG × 1 (RITA-RCA)</td>
<td>Clopidogrel, warfarin</td>
<td>Died of pneumonia</td>
<td>52</td>
</tr>
<tr>
<td>3 2018.12</td>
<td>35/F</td>
<td>Distal descending thoracic aorta (28 mm), distal descending, infrarenal abdominal aorta × 2 (29, 42 mm, sessile)</td>
<td>Spleen, kidney, LE</td>
<td>Hyperhomocysteinemia</td>
<td>33.3</td>
<td>Thoracoabdominal spiral approach, thrombectomy with patch repair × 2</td>
<td>ASA, warfarin (noncompliant)</td>
<td>Thrombus recurrence (medically managed: 2019.4), CI (minor, 2020.9); alive 2021.3</td>
<td>27</td>
</tr>
<tr>
<td>4 2019.3</td>
<td>47/M</td>
<td>Ascending aorta (30 mm, mobile)</td>
<td>CI (major, right MCA)</td>
<td>Factor V Leiden (heterozygous), PE (bilateral PA)</td>
<td>38.6</td>
<td>Sternotomy, thrombectomy with graft replacement, PTE</td>
<td>ASA, warfarin</td>
<td>CTA 2020.10 showing no recurrent thrombus; alive 2022.1</td>
<td>28</td>
</tr>
<tr>
<td>5 2019.7</td>
<td>60/M</td>
<td>Ascending aorta (20 mm, mobile)</td>
<td>Kidney, LE</td>
<td>DVT, PAD with past LE bypass, elevated homocysteine</td>
<td>33.9</td>
<td>Sternotomy, thrombectomy with aortic resection and direct repair</td>
<td>ASA, rivaroxaban</td>
<td>CTA 2021.11 showing no recurrent thrombus; alive 2023.6, lost to cardiac follow-up</td>
<td>26</td>
</tr>
<tr>
<td>6 2019.10</td>
<td>61/F</td>
<td>Ascending aorta (19 mm, mobile)</td>
<td>Bilateral LE</td>
<td>DVT, positive antinuclear antibodies</td>
<td>30.3</td>
<td>Sternotomy, thrombectomy with direct aortic repair, right LE embolectomy and fasciotomies</td>
<td>Warfarin</td>
<td>Alive 2021.9, lost to follow-up thereafter</td>
<td>24</td>
</tr>
<tr>
<td>7 2020.3</td>
<td>57/M</td>
<td>Proximal descending thoracic aorta (28 mm, mobile)</td>
<td>Spleen</td>
<td>Possible lung cancer</td>
<td>28.9</td>
<td>Left thoracotomy, primary repair</td>
<td>ASA, warfarin</td>
<td>Lost to follow-up</td>
<td>1</td>
</tr>
<tr>
<td>8 2020.5</td>
<td>36/F</td>
<td>Ascending aorta (16 mm, mobile)</td>
<td>NSTEMI (LAD)</td>
<td>Substance abuse, decreased protein S and AT III activity, elevated lipoprotein A</td>
<td>34.8</td>
<td>Sternotomy, thrombectomy with direct aortic pair, CABG (LITA-LAD)</td>
<td>ASA, warfarin</td>
<td>Alive 2021.5, lost to follow-up thereafter</td>
<td>12</td>
</tr>
<tr>
<td>9 2020.7</td>
<td>47/M</td>
<td>Distal aortic arch (9 mm, sessile)</td>
<td>CI, kidney, SMA</td>
<td>Antinuclear antibody</td>
<td>34.9</td>
<td>Not operated on for primary aortic thrombus</td>
<td>Apixaban</td>
<td>AKI, ischemic bowel requiring resection for strictures (2020.8); alive 2023.4, no surveillance imaging</td>
<td>3</td>
</tr>
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(Continued)
bilateral pulmonary artery thromboembolectomy was performed at the time of open thrombectomy.

One patient with a sessile PAT at the distal aortic arch (Patient 9), who developed what appeared to be retrograde cerebral infarction, was initially managed pharmacologically. This patient did not undergo open surgery for this PAT because repeat CTA showed that the thrombus had decreased in size in response to anticoagulation, and the patient was discharged to home.

**Postoperative Management and Early Outcomes**

All the patients received IV heparin infusion with a target activated partial prothrombin time of 60 to 80 seconds once postoperative bleeding was controlled. Usually, the patients started an oral anticoagulant, such as warfarin or a novel oral anticoagulant, in addition to aspirin and/or clopidogrel on postoperative day 1, after which IV heparin infusion was discontinued. There were no operative deaths. All patients experienced a steady recovery with no significant complications.

**Patient Follow-up**

During a median follow-up of 18 months (mean, 18.7 ± 15.9 months), 1 patient who had been receiving treatment for HIV infection (patient 2) died of pneumonia related to Coronavirus disease 2019 infection. Two patients experienced recurrence of PAT. One patient (patient 3) was readmitted for chest and abdominal pain at 4 months postoperation and was found to have recurrence of the thrombus in the same location of the distal descending thoracic aorta, extending to the suprarenal and infrarenal abdominal aorta. She was placed on therapeutic IV heparin and was reimaged several days later, with CTA showing a decreased thrombus burden in the aorta. Her symptoms improved, and she was discharged on warfarin with a higher target international normalized ratio because of the recurrence. She was later readmitted to another facility for a new-onset minor cerebral infarction and was then discharged to an acute rehabilitation facility.

Another patient who developed recurrence (patient 10) was not on any oral anticoagulant after surgery because of a previous history of intolerance to oral anticoagulants, and instead was receiving enoxaparin via subcutaneous injection. Two months later, she was readmitted for leg pain and found to have recurrent PAT at the same location in the proximal descending thoracic aorta (Figure 5), as well as an acute right popliteal artery occlusion. She underwent a right femoral artery→posterior tibial artery bypass performed by a vascular surgery team. She presented again at 6 months after the index operation with left upper and lower extremity weakness related to a suspected transient ischemic attack and a significantly worsened descending thoracic aortic thrombus since the prior study. She subsequently underwent a redo left thoracotomy, and the recurrent thrombus was excised. The new thrombus had a larger surface attachment than the original thrombus, as shown in Figure 5. She had a relatively uncomplicated postoperative course and was discharged on postoperative day 11. She was also switched to warfarin with the hope of improving her compliance. Unfortunately, she continued to have poor compliance with anticoagulation and was readmitted for acute mesenteric ischemia secondary to the superior mesenteric artery (SMA) thrombus related to the aortic thrombus recurrence. She required extensive small bowel resection during her last admission, and was ultimately discharged to hospice. See Figure 6 for a graphical abstract of the study.
It is important to note that the patient (patient 9) who did not undergo open surgery for PAT because of evidence of a decrease in size of the original thrombus, was readmitted owing to new onset of abdominal pain. Repeat CTA demonstrated complete disappearance of the original thrombus at the distal aortic arch but showed multiple thrombi at the distal SMA branches and thickened small bowel. The patient underwent small bowel resection secondary to strictures from the small bowel ischemia.

DISCUSSION

PAT is defined as thrombosis in the absence of atherosclerotic or aneurysmal disease of the aorta,1,2 with no cardiac source of the thrombus. This entity is exceedingly rare in clinical practice, with only a few case studies and systemic reviews in the literature. The incidence of aortic thrombus is as low as 0.45% based on autopsy data from >10,000 cadavers in the 1900s3; however, the actual incidence remains to be determined. Owing to the paucity of cases, the management of this condition varies widely and is institution-dependent, with no overriding consensus.

Disease categorization is important, and unlike aortic dissection, there is no universal classification system for PAT. Verma and colleagues1 developed a system in which a thrombus was classified as type I to VI based on its anatomic location: type I, a mural thrombus in the ascending aorta up to the level of the origin of the left subclavian artery (SCA); type II, a thrombus located in the

FIGURE 1. Patient 3. A, Computed tomography angiography (CTA), sagittal view, showing a large thrombus in the distal descending thoracic aorta extending to the suprarenal abdominal aorta. B, CTA, sagittal view, of the abdomen showing multiple large mural thrombi in the abdominal aorta. C, CTA, axial view, showing that approximately one-half of the aortic lumen was obliterated by the mural thrombus, with clear evidence of splenic infarct. Red arrows indicate thrombus as well as splenic infarct.

descending thoracic aorta, between the left SCA and celiac artery; type III, a thrombus located at the level between the celiac artery to the lowest renal artery; and type IV, a thrombus located at the level between the lowest renal artery and the aortic bifurcation. The most common anatomic location of PAT is reportedly the descending thoracic aorta (38%), and the least common is the ascending aorta (12%), as found in the meta-analysis conducted by Fayad and colleagues.4 Interestingly, 5 of our 10 patients presented with a PAT in the aortic root/ascending aorta. The classification system of Verma and colleagues may be useful in determining the optimal management strategy.

The most dreaded complication of an aortic thrombus is embolization to vital end organs.1-4 Unfortunately, many patients already present with this complication at the time of diagnosis. The consequences of cerebral infarction, MI, acute limb ischemia, or mesenteric ischemia can be devastating. The use of anticoagulants with close monitoring and short-interval surveillance imaging should be carefully tailored to the individual patient and thrombus. Features that have been associated with a high risk for embolization and recurrent embolization include pedunculated and highly mobile lesions.5 In the case of a PAT in the ascending aorta or the proximal aortic arch, we prefer urgent open surgery over anticoagulation and surveillance, given the risk of cerebral infarction. In fact, Fayad and colleagues reported in a systemic review that thrombus location in the ascending aorta and the aortic arch were the 2 strongest predictors of embolization recurrence. As for the descending thoracic aorta, the risk of cerebral infarction is low. Although we observed this in 1 patient (patient 9), retrograde embolization to the brain causing cerebral infarction is extremely rare, and this mechanism for stroke has been theorized recently only in severely atherosclerotic aortas but not in those with a PAT. This fact can sometimes allow for a short observation interval with anticoagulation in those who are minimally symptomatic with a descending

thoracic aortic thrombus, as has been shown by some small studies.\textsuperscript{7,8} As demonstrated in 1 of our patients (patient 9), a devastating complication (ischemic bowel) can occur during conservative management even in the course of resolution of the primary PAT. Thus, the conservative decision to anticoagulate and observe should be used cautiously, as there is still risk for distal embolization.

In recent years, TEVAR has become an attractive interventional treatment option because of its less invasiveness, especially for those with descending thoracic aortic aneurysms. This technique can be applied to some patients with PAT in the descending aorta. A literature review by Meyermann and colleagues\textsuperscript{9} found that there is a trend in more recent years to report cases describing successful TEVAR for initial management. Twenty-nine patients (39.2\%) out of 74 patients enrolled in their review with descending PAT initially underwent TEVAR. Twenty-seven (93.1\%) had fully excluded thrombus at the time of the procedure, with no

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recurrence or evidence of repeated embolic phenomena at follow-up. Needless to say, direct manipulation of the PAT during TEVAR can cause significant complication of proximal and/or distal embolization, especially if the thrombus is highly mobile, and the surgical decision should be made carefully among a multidisciplinary team. Also, this technique cannot be applied to a PAT located at the ascending aorta and the aortic arch. Therefore, the open surgical approach should still be considered as the primary treatment in the majority of these cases. One must weigh the risks and benefits of each treatment approach and decide which option is best for each patient.

Out of our 10 patients, 2 had recurrence of the thrombus. This was apparently attributable to nonadherence to anticoagulation. One of our patients (patient 3) had a compliance issue stemming from severe menorrhagia secondary to warfarin; her thrombus resolved during readmission with anticoagulation alone. Another patient (patient 10) had a longstanding history of unprovoked PE on lifelong enoxaparin, for which she had been noncompliant for years before her operation. In the aforementioned literature review, 9 (34.6%) had persistent thrombus, whereas of the 19 patients who initially underwent open surgical repair, 6 (31.6%) had persistent thrombus, 4 of whom underwent subsequent TEVAR. Therefore, continuous careful observation with optimal medical therapy is crucial regardless of the initial treatment to monitor possible recurrence and prevent devastating complications.

Limitations
This study has several limitations. First is its single-center, retrospective observational design with a small sample size, which confers an inherent selection bias. Second, we had no experience using the TEVAR technique in this group of patients, and without a control group, comparison of the results was limited. Third, the duration of follow-up was short, and late results remain unknown.

CONCLUSIONS
This case series demonstrates that open surgical thrombectomy is an excellent treatment modality for primary aortic thrombus in the thoracic aorta. However, careful
postoperative management, including optimal medical therapy, is mandatory to prevent recurrence.

Data Availability Statement
All the coauthors have full access to the patient data, and all the datasets analyzed in the present study are available from the corresponding author on reasonable request.

Conflict of Interest Statement
The authors reported no conflicts of interest.

The Journal policy requires editors and reviewers to disclose conflicts of interest and to decline handling or reviewing manuscripts for which they may have a conflict of interest. The editors and reviewers of this article have no conflicts of interest.

References

Key Words: primary aortic thrombus, distal embolization, open repair, hypercoagulable state