Thrombus in the Non-aneurysmal, Non-atherosclerotic Descending Thoracic Aorta — An Unusual Source of Arterial Embolism

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Abstract  Introduction: Mural thrombus of the thoracic aorta is a rare clinical finding in the absence of aneurysm or atherosclerosis.

Methods: The medical records of all patients diagnosed with a thrombus of a non-aneurysmatic and non-atherosclerotic descending thoracic aorta (NAADTA) and treated by the senior author between 04/1997 and 04/2010 were reviewed.

Results: Eight patients with mural thrombus of the NAADTA were identified. Arterial embolism was the main clinical finding in all cases and involved the lower extremities (n = 6), mesenteric (n = 3) or renal arteries (n = 2). Hypercoagulable disorders were present in 3 cases and a concurrent malignancy in another 3. Two patients underwent open surgery while 4 patients were treated conservatively with anticoagulation. Of the remaining 2 patients, one was treated with a thoracic stent-graft and aorto-biiliac bypass and the other one with transfemoral thrombectomy. Technical success was achieved in all surgical cases and thrombus resolution or stable disease in the conservative management group. No thrombus recurrence was observed during a mean follow-up of 49 months.

Conclusion: The management of mural thrombus in NAADTA represents a challenge, especially in case of malignant disease or hypercoagulable disorder as a potential underlying pathology.
Introduction

Distal arterial embolism is a relatively common problem that carries increased morbidity and, potentially, mortality. The amputation rate following acute limb ischaemia is estimated at 13–14% while mortality is at 9–12%. Over 80% of all peripheral and visceral emboli originate from disturbances of cardiac function itself such as atrial fibrillation, myocardial infarction, endocarditis and prosthetic heart valves. Non-cardiac causes include aortic pathologies such as aneurysmal lesions, dissections, penetrating ulcers or traumatic lesions.

Since 1967 when Oliver et al. published the first described case of thrombo-embolism from the thoracic aorta, few case reports and small series of patients with thrombus in a non-aneurysmal thoracic aorta have been published. In most of these cases, peripheral embolism was the initial clinical manifestation. The expanding availability of advanced diagnostic modalities such as computed tomography (CT), trans-esophageal echocardiography (TEE) and magnetic resonance imaging (MRI) increasingly made possible the accurate diagnosis of this aortic pathology over the past decade.

The optimal management of these patients is still controversial and depends on the location and morphology of the thrombus, the symptoms and the general condition of the patient. The presence of a floating thrombus in the thoracic aorta is associated with a high risk of peripheral embolism. The primary objective of this study is to report our experience regarding the diagnosis and management of thrombus in the non-aneurysmal, non-atherosclerotic descending thoracic aorta (NAADTA).

Patients and Methods

The medical records of all patients diagnosed with thrombus in an NAADTA and treated by the senior author were reviewed retrospectively. For the descending thoracic aorta, aneurysmal dilatation was defined as an aorta at least twice the diameter of the patient’s contiguous normal aortic calibre. The presence of atherosclerosis was graded based on the amount of calcification identified on the CT scan. Data collection included patient demographics, risk factors for vascular disease, symptoms leading to diagnosis, relevant co-morbidities, diagnostic and therapeutic management and outcome. Treatment complications and the follow-up outpatient-clinic evaluations were reviewed. All imaging studies were evaluated independently by vascular radiologists.

Results

In a period of 12 years, 8 patients (7 female, 1 male) were identified with thrombus in an NAADTA. The mean age of the patients was 55 years (range, 45–68). Further demographic data and clinical findings of the patients are summarised in Table 1.

Diagnostic management

The diagnostic work-up that revealed intraluminal aortic thrombus was prompted by lower extremity embolism in 6 cases and mesenteric embolism in 1 case. In an additional case, it was an incidental finding in a staging CT scan for malignant melanoma. Imaging modalities included multi-detector CT scan alone or in conjunction with magnetic resonance angiography (MRA) and TEE in all cases, while digital subtraction angiography (DSA) was performed on demand. Screening for hypercoagulable disorders was performed in all cases prior to initiation of treatment.

The thrombus was localised in zone III of the thoracic aorta in 5 patients and extended to the visceral abdominal aorta in three cases. The sites of arterial embolism involved primarily the lower extremity arteries and, less frequently, the visceral arteries (Table 1).

Characteristics of aortic lesions — underlying pathologies

None of the patients had concomitant aortic or cardiac pathologies such as aneurysm, penetrating aortic ulcer, severe atherosclerosis or calcification of the thoracic aortic wall or intracardial thrombus. One patient had an aortic wall angiosarcoma, which was associated with mural thrombus formation. The abdominal aorta, as well as the iliac arteries, was free of aneurysmal disease or other pathology, which could account for peripheral embolism. Minimal calcification of the abdominal aorta was present in 3 out of 8 patients.

The screening for hypercoagulable disorders revealed anti-phospholipid syndrome in 1 patient and heparin-induced thrombocytopenia (HIT) type II in another. Although not diagnosed primarily, one patient underwent repeated extensive work-up for thrombophilia during follow-up, which eventually revealed elevated serum levels of plasminogen activator inhibitor-1 (PAI-1), positive antinuclear antibodies (ANAs) and anti-neutrophil cytoplasmic antibodies (ANCAs), suggesting the diagnosis of sub-clinical vasculitis. Concurrent malignancies were present in three patients and included stage IV malignant melanoma, myeloproliferative malignancy and intimal sarcoma of the thoracic aorta.

The thrombus was sessile in 5 cases and pedicled in 3.

Therapeutic management

Four patients were treated conservatively. Two were placed on intravenous heparin followed by oral anticoagulation with coumadin derivatives, while one patient with Rutherford class I chronic limb ischaemia due to...
chronic occlusion of the left iliac artery and stage IV malignant melanoma was only given Aspirin, due to the advanced tumour disease and the stability of the mural thrombus. The choice of conservative treatment was based on the presence of significant co-morbidities in the first patient (Fig. 1(a)–(c)) and advanced malignancy in the latter patient. The third patient had a known anti-phospholipid syndrome and a sessile thrombus of the thoracic aorta. The fourth patient had been therapeutically hep-arinised during the thromboembolectomy of the lower extremities and maintained on anticoagulation thereafter. However, a four-compartment fasciotomy was eventually necessary. Follow-up CT scan 2 months later showed complete resolution of the aortic thrombus.

Surgical treatment was pursued in 4 patients. One patient at high risk for thoracotomy underwent a less-invasive procedure with transfemoral thrombectomy with a Fogarty catheter under fluoroscopic guidance and bilateral femoral thromboembolectomies, recognising the risk of visceral or renal embolisation. Due to unrecognised mesenteric embolism during the procedure, open surgery to remove embolic material from the superior mesenteric artery was necessary a day later.

The second patient with intimal sarcoma of the thoracic aortic wall and liver metastases underwent palliative excision of the aortic tumour and direct closure of the aorta. The second patient with intimal sarcoma of the thoracic aortic wall and liver metastases underwent palliative excision of the aortic tumour and direct closure of the aorta.11 In the third patient of that group, an aortic tumour was suspected on CT scan. To achieve complete tumour resection, a segmental resection of the descending thoracic aorta and interposition of a Dacron graft was performed after establishment of an arterio-arterial extracorporeal circulation with the BioMedicus pump (Medtronic, Eden Prairie, MN, USA). However, malignancy was not confirmed on histopathologic examination. At the 10-year follow-up after aortic replacement, the patient underwent repeat extensive work-up for thrombophilia, which eventually revealed elevated values of PAI-1. The last patient treated surgically had extensive thrombus in the descending thoracic and pararenal aorta and a chronic infrarenal aortic occlusion, possibly due to prior embolic

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**Table 1** Demographic and clinical data of all 8 patients with a thrombus of a non-aneurysmal and non-atherosclerotic descending thoracic aorta.

<table>
<thead>
<tr>
<th>Patient (gender, age)</th>
<th>Localization of emboli</th>
<th>Relevant concomitant disease</th>
<th>Diagnosis</th>
<th>Therapy</th>
<th>FU (ms)</th>
<th>Result</th>
</tr>
</thead>
<tbody>
<tr>
<td>No. 1 (female, 45 years)</td>
<td>Bilateral common femoral arteries, superior mesenteric artery</td>
<td>Heparin-induced thrombocytopenia type II</td>
<td>TEE, DSA, CT</td>
<td>Fogarty manoeuvre, embolectomy, Coumadin derivatives</td>
<td>122</td>
<td>No recurrence</td>
</tr>
<tr>
<td>No. 2 (female, 64 years)</td>
<td>Bilateral popliteal and crural arteries</td>
<td>Aortic sarcoma</td>
<td>TEE, MRA, CT</td>
<td>Resection, Aspirin</td>
<td>18</td>
<td>Died from metastases</td>
</tr>
<tr>
<td>No. 3 (female, 56 years)</td>
<td>Bilateral common femoral arteries, deep femoral and popliteal arteries</td>
<td>Unknown</td>
<td>TEE, CT</td>
<td>Conservative, Coumadin derivatives</td>
<td>20 (lost to FU)</td>
<td>No recurrence</td>
</tr>
<tr>
<td>No. 4 (female, 51 years)</td>
<td>Mesenteric arteries</td>
<td>Elevated values of plasminogen activator inhibitor-1</td>
<td>TEE, MRA, CT</td>
<td>Resection, aortic replacement, Coumadin derivatives</td>
<td>88</td>
<td>No recurrence</td>
</tr>
<tr>
<td>No. 5 (female, 68 years)</td>
<td>Right common iliac artery and common femoral artery, right renal artery, celiac trunk</td>
<td>Antiphospholipid syndrome</td>
<td>TEE, CT</td>
<td>Conservative, Coumadin derivatives</td>
<td>78</td>
<td>No recurrence</td>
</tr>
<tr>
<td>No. 6 (female, 53 years)</td>
<td>Infrarenal aorta, bilateral common and external iliac arteries</td>
<td>Myeloproliferative malignancy-essential thrombocytosis</td>
<td>TEE, DSA, CT</td>
<td>Complex repair incl. stent-graft and aortobifemoral bypass</td>
<td>51</td>
<td>No recurrence</td>
</tr>
<tr>
<td>No. 7 (male, 57 years)</td>
<td>Left common iliac artery, right femoral artery</td>
<td>Malignant melanoma</td>
<td>TEE, CT</td>
<td>Conservative, Aspirin (due to advanced metastatic disease)</td>
<td>12</td>
<td>Persistent thrombus, no progression</td>
</tr>
<tr>
<td>No. 8 (female, 49 years)</td>
<td>Bilateral popliteal and crural arteries</td>
<td>Unknown</td>
<td>TEE, CT</td>
<td>Conservative, Aspirin (no compliance for Coumadin derivatives)</td>
<td>3</td>
<td>Thrombus resolution</td>
</tr>
</tbody>
</table>

FU, follow-up; ms, months; DSA, digital subtraction angiography; TEE, trans-esophageal echocardiography; MRA, magnetic resonance angiography; CT, computed tomography.
events. The occlusive disease was associated with a Ruther-
der class III chronic limb ischaemia. The initial opera-
tion included an aortobifemoral bypass after transaortic
balloon thrombectomy of the thoracic aorta with tempo-
rary occlusion of the visceral vessels. Due to the presence
of residual thrombus in the thoracic aorta as demonstrated
on the postoperative CT scan, a transfemoral thoracic
stent-graft (Zenith, TX1, 30/80 mm William Cook Europe,
Bjaerverskov, Denmark) was used to cover the involved
segment of the thoracic aorta and prevent further embo-
larism (Fig. 2(a)–(c)).

Outcomes and follow-up

Follow-up for both surgical and conservative treatment
groups included regular office visits and thoraco-abdominal
CT scans. CT scans were performed every 6 months for the
first 2 years and individually thereafter.

The intraluminal thrombus had completely resolved in
both patients treated with coumadin derivatives, and no
recurrence was observed on follow-up imaging. One
patient with anti-phospholipid syndrome was maintained
on lifelong anticoagulation, while the other patient in the
conservative group was switched to Aspirin 12 months
later following an upper gastrointestinal bleed. The latter
patient was lost to follow-up after 20 months. The
patient with malignant melanoma treated with Aspirin
alone showed no progression of the initially sessile,
circumferential mural thrombus and there were no
further signs of peripheral embolism. Therefore, conser-
ervative therapy was continued indefinitely. The fourth
patient treated conservatively, was given Aspirin therapy
lifelong due to poor compliance with coumadin
derivatives.

The postoperative course of the surgically treated
patients was uneventful and no adverse event or thrombus
recurrence was diagnosed during follow-up. Anticoagulation
in the surgically treated group of patients included coumadin
derivatives in 2 patients. One patient was put on Aspirin after
resection of the aortic sarcoma, which had caused the
thrombus, while the second patient declined oral anti-
coagulation with coumadin derivatives. The patient with
aortic sarcoma died of generalised metastases 18 months
after the operation.
Discussion

The incidence of aortic mural thrombus was reported by Machleder et al.\textsuperscript{12} to be as low as 0.9% (95 cases) in a study of 10,671 autopsies. Only about half of these patients had aneurysmal disease and only 17% of the patients had evidence of peripheral embolism. The authors described the nature of the thrombus as a distinct entity in many cases. During the same time period, only 2 cases of symptomatic aortic mural thrombus were treated at that institution.

The true incidence of mural thrombus in NAADTAs is unknown but it is possibly much higher than that reported in the literature.

The reported cases of mobile thrombus in the thoracic aorta were limited to 100 published cases in a review by Choukroun et al.\textsuperscript{10} In an additional review of the literature since 2002, we were able to identify about 50 more cases, including a few small series.\textsuperscript{13–19}

In most cases, the thrombus was located in the descending thoracic aorta, although the presence of thrombus in the aortic arch as well as in the abdominal aorta has been described.\textsuperscript{10,20}

The aetiology of thrombus formation in a macroscopically normal aorta is not well understood. A correlation with underlying malignant disease, hypercoagulable disorders, primary endothelial disorders or even iatrogenic causes has been suggested (Table 2).\textsuperscript{7,18,20,21–24} Advanced but not early atherosclerotic lesions can be identified on CT images from the presence of calcifications and aortic wall irregularities. The presence of tumour in the thoracic aorta could be a predisposing factor for thrombus formation. Most aortic sarcomas were diagnosed following aortic thrombus formation complicated with peripheral or visceral embolism.\textsuperscript{25,26}

Similarly, in our series, a suspicious intraluminal growth proved to be an aortic sarcoma. In patients with aortic mural thrombus, it is very important to evaluate the aortic wall thoroughly and consider always the possibility of a local malignancy. However, the differential diagnosis between mural thrombus with or without aortic wall malignancy can be difficult in certain cases (patient No. 4, Table 1).

In the present series, 6 out of 8 patients had a concurrent hypercoagulable state as a possible underlying mechanism.

Several publications\textsuperscript{8,18} have demonstrated that genetic and thrombophilic factors could be associated with thrombus formation in the NAADTAs. Recent advances in genetic diagnosis have shown a correlation between certain genetic patterns and thrombus formation at other vascular sites.\textsuperscript{27} A thorough genetic and hypercoagulability work-up should be performed in all patients with thrombus in the NAADTAs. It is possible that, in the earlier years of this series, some patients were not screened to today’s standards due to technical limitations at that time.

Thrombus formation in the thoracic aorta is encountered with increasing frequency over the past 10 years. This is mainly through the use of advanced imaging as part of thorough work-up to identify the source of peripheral or visceral embolism. In most centres, standard postoperative evaluation of patients with peripheral embolism includes 24-h electrocardiography (ECG), TEE, CT angiography or MRI of the thoraco-abdominal aorta and arterial duplex ultrasound of the vessels proximal to the embolic occlusion.

Several authors have earlier reported TEE to be an adequate method for detecting mural thrombus of the thoracic aorta,\textsuperscript{9} with some suggesting TEE to be comparatively more sensitive than CT or MRI.\textsuperscript{28} However, these studies were published before recent advancements in CT and MR imaging with high-resolution scanners.

Of the 8 patients in our series, 7 were diagnosed during the work-up for lower extremity or mesenteric ischaemia. In general, mural thoracic aortic thrombus is rarely diagnosed before becoming symptomatic.

The definitions regarding thrombus material in the thoracic aorta are relatively unclear because no consistent nomenclature exists and the terms ‘mobile’, ‘floating’, ‘sessile’ or ‘pedicled’ were, in all cases, defined by the treating physicians. Due to the limited number of cases, the optimal treatment is often debatable and different approaches have been suggested. Several authors\textsuperscript{29,30} have described catheter-guided thrombolysis to treat mural aortic thrombus, a therapy that has later been associated with a significant risk of additional peripheral embolism. In particular, there is the potential danger of lysing pedunculated lesions, releasing the bulk of the thrombus and causing massive embolisation.\textsuperscript{30}

Other series have demonstrated that therapeutic oral anticoagulation is an efficient and safe option in patients with aortic mural thrombus.\textsuperscript{20,31} However, a careful patient selection is necessary, as cases of persistent or recurrent embolus under coumadin derivatives have been described.\textsuperscript{32} The proposed duration of oral anticoagulation varies among authors, ranging in time from complete resolution of the thrombus to lifelong. In a review of patient outcomes in 23 cases, complete thrombus resolution occurred in 74% (n = 17) of the cases.\textsuperscript{6,7,10,31,33–35}

In one of the largest series reported so far, Choukroun et al.\textsuperscript{10} suggested that surgical treatment should be

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**Table 2.** Underlying pathologies described for the presence of thrombus in the non-aneurysmal, non-atherosclerotic thoracic aorta.

<table>
<thead>
<tr>
<th>Predisposing factor</th>
<th>References</th>
</tr>
</thead>
<tbody>
<tr>
<td>Essential thrombocytopenia\textsuperscript{7,35,43}</td>
<td></td>
</tr>
<tr>
<td>Protein C or S deficiency\textsuperscript{7,18,20,44,45}</td>
<td></td>
</tr>
<tr>
<td>Antiphospholipid syndrome\textsuperscript{23,46,47}</td>
<td></td>
</tr>
<tr>
<td>Heparin-induced thrombocytopenia\textsuperscript{a,b} 48,49</td>
<td></td>
</tr>
<tr>
<td>Hyperhomocysteinemia\textsuperscript{15,20}</td>
<td></td>
</tr>
<tr>
<td>Iatrogenic (e.g. intraaortic balloon pumping)\textsuperscript{50,22}</td>
<td></td>
</tr>
<tr>
<td>Aortic wall tumor\textsuperscript{51}</td>
<td></td>
</tr>
<tr>
<td>Cancer\textsuperscript{20,24,44,47,52}</td>
<td></td>
</tr>
<tr>
<td>Chemotherapy\textsuperscript{33,54}</td>
<td></td>
</tr>
<tr>
<td>Homozygous plasminogen activator inhibitor type 1 (PAI-1): 675(4G) and heterozygous GP Ia 807C/T genotypes\textsuperscript{6}</td>
<td></td>
</tr>
<tr>
<td>Factor V Leiden\textsuperscript{55}</td>
<td></td>
</tr>
<tr>
<td>Familial dysfibrinogenemia\textsuperscript{a}</td>
<td></td>
</tr>
<tr>
<td>Antithrombin III deficiency\textsuperscript{20}</td>
<td></td>
</tr>
<tr>
<td>Blunt aortic trauma\textsuperscript{19,21,56}</td>
<td></td>
</tr>
<tr>
<td>Drug abuse\textsuperscript{4}</td>
<td></td>
</tr>
<tr>
<td>Elevated Factor VIII\textsuperscript{57}</td>
<td></td>
</tr>
<tr>
<td>Aspergiloma\textsuperscript{58}</td>
<td></td>
</tr>
<tr>
<td>Rheumatic disease\textsuperscript{59,60}</td>
<td></td>
</tr>
</tbody>
</table>
reserved for patients not responding with thrombus resolution on follow-up TEE after 2 weeks of therapeutic anticoagulation with heparin. In this series as well as in earlier series, surgical treatment included thrombectomy and segmental aortic resection. Segmental aortic resection provides the optimal treatment in patients with neoplasms of the thoracic aorta as an underlying cause for thrombus formation. In these cases, en-bloc resection with negative margins should invariably be attempted, unless limited by extensive metastatic disease.

Our literature review identified 8 cases of patients with thrombus of the thoracic aorta treated with stent-grafts. A favourable outcome without complications was described in all cases. However, the risk of distal embolism during wire manipulation and stent-graft deployment could represent a limitation of endovascular treatment. Nevertheless, no such complications were described in the reported cases. Full entrapment of the thrombus between the stent-graft and the aortic wall should be achieved. Therefore, the required length of the graft should be at least 2 cm proximally to 2 cm distally to the longitudinal extension of the thrombus. We preferred an oversizing of the stent-graft by 5–10%.

In general, no clear guidelines exist regarding the optimal management of these patients. Possible reasons, therefore, include the rarity of the disease, the limited available experience and the protean pathophysiology. Most authors would treat patients with sessile aortic wall thrombus using intravenous heparin followed by oral therapeutic anticoagulation. However, the choice of patients requiring surgery and what is the best surgical approach remain controversial issues.

In our clinical practice, all patients are placed initially on intravenous heparin anticoagulation. Factors determining subsequent management include the patient’s co-morbidities, thrombus size and morphology, current symptoms and risk factors predisposing to thrombus formation. Patients with concurrent malignancy, coagulation disorders and sessile thrombi are preferably treated conservatively.

When a mobile thrombus is encountered or recurrent episodes of embolism from a sessile thrombus despite therapeutic anticoagulation occur, we advocate an endovascular approach using a stent-graft to fix the thrombus to the aortic wall. We reserve open repair for cases not suitable for endoluminal treatment or for patients with suspected malignancy.

One of the limitations of this retrospective study is that it covers a time period of 12 years with current diagnostic and therapeutic modalities not being available in the earlier cases.

Conclusion

Thrombus in the descending, otherwise ‘healthy’, thoracic aorta represents an underdiagnosed medical entity, which could potentially explain many cases of cryptogenic embolism. An underlying cause should be thoroughly searched for, including thrombophilia studies. Aortic wall tumour as well as malignancy elsewhere should be excluded. Currently, long-term anticoagulation is the most widely accepted first-line therapy. Surgical intervention is reserved for patients with contraindications to long-term anticoagulation or those who fail conservative management. We consider the presence of a mobile thrombus as a relative indication for primary intervention due to the high risk of embolism. Transaortic or transfemoral thrombectomy, segmental aortic resection and stent-graft implantation are among the surgical options with the latter gaining preference in our institution.

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Conflict of Interest
No.

Ethical Approval
N/A.

References


Embolism from Thrombus in the Normal Aorta


