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# Recurrent lower limb embolism from thoracic aortic mural thrombus: a rare presentation of occult malignancy

## 因胸腔主動脈附壁血栓引致的復發性下肢栓塞：隱匿性癌症的一個罕有表徵

Initial presentation of a malignant disease as recurrent attacks of lower limb ischaemia due to emboli from a mural thrombus in the descending thoracic aorta is extremely rare. A diagnosis of malignancy may thus easily be overlooked. Recent advances in imaging technology have made the diagnosis of thoracic aortic mural thrombi much easier. Occult malignancy should always be suspected in the absence of biochemical evidence of hypercoagulability. We report on a patient with underlying malignant disease who presented with lower limb ischaemia that was relieved by axillobifemoral bypass.

癌症的初步表徵，很少會出現主動脈附壁血栓引起的下肢缺血，因此診斷時很容易忽略了患癌症的可能。近年造影技術的進步令診斷胸腔主動脈附壁血栓比以前容易。在沒有出現過度凝血的情況，便要考慮是否患有隱匿性癌症。本文報告一名出現下肢缺血表徵的隱匿性癌症患者，其病症在接受腋下至股部動脈搭橋手術後得到舒緩。

### Key words:

Aortic diseases;  
 Embolism;  
 Magnetic resonance imaging;  
 Thrombosis;  
 Tomography, X-ray computed

### 關鍵詞：

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 栓塞；  
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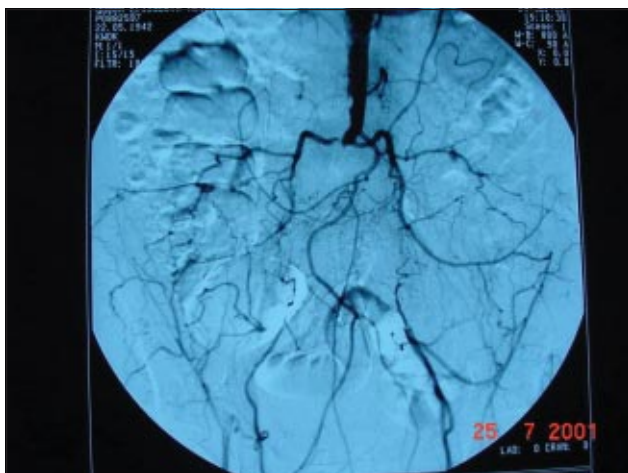
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### Introduction

Cardiac mural thrombus and atheromatous plaques are the most common origins of arterial emboli and account for approximately 85% of all thromboembolic events.<sup>1</sup> The remaining 15% is considered 'cryptogenic'. Recent advanced imaging technology has revealed that, although rare, some of these 'cryptogenic' emboli are caused by an aortic mural thrombus within a normal aorta. Studies have shown that it actually accounts for 5% to 9% of peripheral embolisation.<sup>2,3</sup> Nonetheless recurrent attacks of peripheral embolism from a thoracic aortic mural thrombus (TAMT) as the initial presentation of underlying occult malignancy is seldom reported. Clinicians unaware of this rare vascular presentation may easily overlook the TAMT and fail to diagnose the underlying malignancy.

### Case report

A 57-year-old Japanese man presented in February 2000 with sudden-onset bilateral lower limb pain. He was a chronic smoker who had good past health. Physical examination revealed pale and cool feet with slow capillary return but intact motor and sensory functions. The femoral pulses were normal but all distal pulses were absent. The ankle brachial index (ABI) was 0.19 on the right side and 0.53 on the left. Urgent lower limb



**Fig 1. Angiogram showing complete occlusion of distal abdominal aorta with extensive collaterals**



**Fig 2. Thoracic aortogram showing luminal filling defect at thoracic aorta just distal to left subclavian artery**

angiogram demonstrated occlusion of the right distal superficial femoral, bilateral popliteal, bilateral trifurcation, and proximal peroneal arteries with intraluminal filling defects suggestive of emboli. Other vessels, including the abdominal aorta and its proximal branches, were normal with no features suggestive of atherosclerosis. The patient was diagnosed with acute ischaemic limb due to embolism. Intra-arterial thrombolysis was performed with 2 million units of intravenous urokinase, and thrombi in the right superficial femoral artery and bilateral popliteal arteries were dissolved. Some residue thrombi remained in the crural vessels. Ischaemia improved dramatically after thrombolysis and no further interventions were deemed necessary. Echocardiogram and transoesophageal ultrasound to identify the source of emboli were normal. The patient was discharged with a 6-month course of warfarin. He had a mild degree of claudication in both legs that was well-tolerated at subsequent follow-up.

He remained well until 10 months later when he developed increasing claudication in his left leg for 1 week. The ABI was 0.52 on the right side and 0.4 on the left. A further set of lower limb angiograms revealed increasing arterial occlusion affecting both popliteal and crural arteries with extensive collateral supply. The patient opted for conservative treatment as he could tolerate the symptoms well. Blood tests for hypercoagulability were again normal.

He defaulted from further out-patient follow-up until 6 months later when he developed acute onset of bilateral lower limb rest pain with features of acute ischaemia. All lower limb pulses were absent with

unrecordable ABIs. Surprisingly, a 1-cm immobile firm-to-hard left supraclavicular lymph node was identified. Urgent lower limb arteriogram and aortogram showed complete occlusion of the bilateral common iliac and external iliac arteries and lower abdominal aorta from L4 downward with extensive collateral from the lumbar arteries (Fig 1). The distal arteries of the leg were unchanged. Thoracic aortogram was obtained in view of the atypical presentation and showed a large filling defect in the descending thoracic aorta just below the left subclavian artery (Fig 2). In view of the enlarged supraclavicular lymph node, a diagnosis of recurrent peripheral emboli due to TAMT, possibly related to an underlying malignancy, was finally made. Emergency embolectomy failed and the lower limb was revascularised by means of a right axillobifemoral bypass using a 10-mm external ring-supported Dacron graft. Postoperatively his rest pain was relieved but section and histology of the femoral artery thrombus showed organised thrombus indicative of significant chronicity. There were no tumour cells identified within the thrombus. Fine needle aspiration of the cervical lymph node showed metastatic adenocarcinoma. Further computed tomography (CT) and magnetic resonance imaging (MRI) of the thorax confirmed a mural thrombus in the descending thoracic aorta with multiple para-aortic lymphadenopathy. There was no evidence of direct invasion of tumour mass into the aorta and there were no mass lesions in the lungs nor hilar or mediastinal lymphadenopathy. A diagnosis of metastatic carcinoma of unknown origin was confirmed.

As the patient had no other symptoms, he refused

further invasive investigations to identify the primary tumour. He was assessed by a clinical oncologist and again refused further therapy for the metastatic lymph nodes. He made a satisfactory postoperative recovery, was able to walk unaided, and was discharged on long-term warfarin after 2 weeks. He lived for another 9 months with no further ischaemic symptoms. Prior to death he developed acute renal failure and CT showed intra-abdominal lymphadenopathy and mildly dilated renal pelvises and proximal ureters. This may have been secondary to extrinsic compression from the lymph nodes or vascular thrombosis. His symptoms were compatible with obstructive uropathy.

## Discussion

Thoracic aortic mural thrombus in an otherwise normal aorta as a cause of peripheral embolisation was first described in 1981.<sup>4</sup> Later studies and a large autopsy series also described TAMT in the aorta with no aneurysm and intimal disease.<sup>2,3,5</sup> These series clearly established that TAMT should be considered a potential source of emboli when investigation of atheromatous and cardiac sources are inconclusive.

Diagnosis of TAMT requires a high level of clinical suspicion and availability of advanced imaging technology. The availability and use of transoesophageal echocardiogram (TEE), CT scan, and MRI have revealed that TAMT may account for up to 9% of all cases of arterial thromboembolism.<sup>3</sup> Since TAMT represents an important source of arterial thromboembolism that has been previously underdiagnosed, it is now suggested that a thorough diagnostic workup of the aorta, preferably by means of CT and MRI, should be performed in all patients in whom other sources of embolisation have been excluded.<sup>3</sup> Magnetic resonance imaging and spiral CT are more sensitive than TEE in detecting TAMT.<sup>6</sup>

The pathogenesis of TAMT formation has not been defined. A number of studies have shown that generalised hypercoagulability is an important prerequisite for the development of TAMT.<sup>7-10</sup> The associated circulating factor abnormalities or antibodies include antiphospholipid antibodies,<sup>11</sup> activated protein C deficiency,<sup>3</sup> anticardiolipin antibodies,<sup>8</sup> antithrombin III deficiency,<sup>12</sup> protein C and S deficiencies,<sup>13</sup> elevated factor VIII and familial dysfibrinogenemia.<sup>7</sup> Potential protagonists of thrombosis include hyperhomocysteinaemia, polycythaemia rubra vera, and malignancy.<sup>7</sup> It is thus vital that all patients with TAMT undergo biochemical screening for hypercoagulability. In the absence of any positive

findings, underlying occult malignancy should be suspected.

With hindsight, one might conclude that the patient in this case review had an occult malignancy that induced a state of generalised hypercoagulability that in turn resulted in the formation of TAMT. It was unfortunate that his first clinical presentation was of acute lower limb ischaemia due to emboli from the TAMT, an extremely rare presentation and the significance of which we were unaware of. The TAMT finally resulted in recurrent attacks of lower limb embolism and the malignancy spread to the supraclavicular lymph nodes.

By the time the diagnosis was made the malignancy was too advanced for definitive treatment. Palliative treatment was prescribed in the form of axillobifemoral bypass to relieve limb ischaemia and long-term anticoagulant therapy to prevent further thromboembolism.

We concur with a number of studies that all patients with acute limb ischaemia in the absence of any identifiable cardiac source of embolism should undergo TEE, CT scan, or MRI to examine the aorta.<sup>3,5,6</sup> When TAMT is identified, the underlying pathogenesis should always be sought by performing biochemical tests for hypercoagulation disorders. When no cause for the TAMT can be identified, the possibility of occult malignancy should be suspected and further investigations performed, for example endoscopy.

There is no consensus on the optimal treatment for TAMT and each case must be judged individually. Treatment with thrombolysis,<sup>14</sup> open thrombectomy,<sup>2-4</sup> aortic stenting, and systemic anticoagulation<sup>7</sup> have all been successful. Systemic anticoagulation appears to be the least invasive and effectively prevents further thromboembolism. In this patient with a limited life expectancy due to disseminated malignancy, palliative axillobifemoral bypass to relieve limb ischaemia due to a blocked aorta was the most reasonable therapy. Long-term anticoagulation also prevented further thromboembolic events.

## Conclusion

Thoracic aortic mural thrombus is a rare cause of acute lower limb ischaemia. As the initial presenting symptom of occult malignancy is extremely rare, a high level of clinical suspicion is necessary to make the diagnosis. Patients with unidentifiable cardiac or atheromatous source of peripheral emboli undergo

thorough examination of the aorta, preferably with CT and MRI. Occult malignancy as the cause of TAMT should always be considered, especially when biochemical tests for hypercoagulability are negative. All clinicians should be aware of this uncommon vascular presentation in order to make an early diagnosis.

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