CASE REPORT

Large aortic arch mural thrombus in non-atherosclerotic thoracic aorta—a rare cause of stroke in the young

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Accepted 3 November 2015

SUMMARY

A 43-year-old Indian woman was admitted to the intensive care unit, with large cerebellar infarction. A brain CT scan showed a large non-haemorrhagic infarct involving the vermis and cerebellar hemispheres. Transesophageal echocardiography and CT angiography demonstrated a large mural aortic thrombus. No evidence of cardiac or vascular pathology was present. The patient had a high risk for surgical intervention and was treated with anticoagulation. Follow-up imaging 2 weeks later showed complete resolution of the thoracic aortic thrombus. This case report emphasises that imaging of the great vessels of the neck and transesophageal echocardiography should be included in ischaemic stroke work up. A thoracic aortic thrombus should always be considered in young patients with unexplained stroke or peripheral embolism; the condition can be treated effectively with anticoagulation.

BACKGROUND

A mural thrombus in the aortic arch is a very rare cause of systemic embolism and often goes undiagnosed. The optimal management for this condition remains controversial. This patient had a large mural aortic arch thrombus in the non-atherosclerotic aorta, which was treated with anticoagulant. This treatment led to a favourable outcome.

CASE PRESENTATION

A 43-year-old Indian woman with no medical history of note presented to the emergency department of the Ahmadi Hospital, with a sudden onset of left-sided hemiparesis accompanied by cerebellar signs. She had no family history of hypercoagulability.

INVESTIGATIONS

The patient was admitted to the intensive care unit. An urgent brain CT scan showed a large non-haemorrhagic infarct involving the vermis and cerebellar hemispheres (figure 1). She also had a weak pulse in the left upper limb and arterial Doppler demonstrated ischaemia.

ECG and cardiac monitoring showed normal sinus rhythm. No evidence of general cardiac pathology or valvular heart disease was evident, and the foramen ovale was not patent.

CT angiography for the aorta and large vessels of the neck showed a mass in the aortic arch (thrombus vs angiosarcoma) (figure 2). The entire thoracic aorta had normal dimensions with no visible

Figure 1 Large non-haemorrhagic infarction involving the vermis and cerebellar hemispheres.

atherosclerosis. Transesophageal echocardiography (TEE) revealed an echo-dense mass (figure 3), measuring 12 mm in maximum diameter, directly attached to the aorta. No evidence of underlying atherosclerotic disease was present. The patient was not dehydrated. She had no family history of hypercoagulability. She was not on hormone therapy and was not using oral contraceptives. Her vascular risk factors were negative for hypertension, diabetes mellitus, cigarette smoking and hypercholesterolaemia. Haemoglobin was normal and testes were negative for antinuclear antibody,

Figure 2 Aortic arch mass (arrows) on CT of the chest, with contrast.

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To cite: Aldaher MK, El-Fayed HM, Malouka SA, et al. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2015-212887

Figure 3 Transesophageal echocardiography showing a large non-calcified verrucous mass (arrows) attached directly to the aortic wall, without evidence of atherosclerotic disease.

antineutrophil cytoplasmic antibody, anti-DNA and anti-Cardiolipin IgG and IgM.

DIFFERENTIAL DIAGNOSIS

The differential diagnosis was thrombus due to vasculitis, a hypercoagulability state such as polycythaemia vera, depressed activation of protein C, protein S deficiency, antiphospholipid antibody, atherosclerotic disease or angiosarcoma.

Vasculitis was ruled out by negative immunology studies. The patient was not dehydrated, and her haematocrit was normal. A thrombophilia screen was not initially performed due to early treatment with anticoagulation ischaemia in the left upper limb.

TREATMENT

The high operative risk led to a decision to treat the case as thrombus using anticoagulation. The patient was given intravenous heparin and later transitioned to warfarin with a target international normalised ratio (INR) of 2–3.

OUTCOME AND FOLLOW-UP

An early follow-up with MR aortography (figure 4) was performed 2 weeks later due to the possibility of angiosarcoma.

Figure 4 Follow-up MR angiography of the thoracic aorta showing complete resolution of the mass.

The imaging showed complete resolution of the thoracic aorta mass. The patient's neurological status continued to improve and she remains on warfarin.

The decision was made to keep the patient on warfarin with a target INR of 2–3 for 3 months, and re-evaluate thereafter for thrombophilia.

DISCUSSION

Atherosclerotic plaque-related thoracic aortic thrombus is frequently experienced by elderly patients and is usually associated with diffuse atherosclerotic disease.^{1–3} Thrombus within the thoracic aorta in the absence of underlying atherosclerosis or aortic aneurysm is very rare. Evidence of this condition is limited to a few small series or case reports.^{1–6} Choukroun *et al*¹ reported their findings on nine patients with mobile thrombus of the thoracic aorta. Thrombus, either sessile or pedunculated and mobile, was detected during work up for a peripheral or cerebral embolic event. No evidence of atheromatous disease was present at the thrombus attachment site in three patients. One patient required surgery because of persistent thrombus despite anticoagulation. The thrombus was in the aortic arch in one patient. Tsilimparis *et al*² studied eight patients with thrombus in the non-aneurysmal non-atherosclerotic descending thoracic aorta on imaging. Seven of the eight patients were female and the mean age was 55 years (range 45–68). Six patients had a concurrent hypercoagulability state and three had concurrent malignancies. Three patients were treated conservatively with warfarin and thrombus resolution occurred.

Laperche *et al*³ studied 23 young patients (mean age 45 years; 10 women) with mobile aortic arch thrombus, from a total of 27855 TEE examinations. All but three had one or more cardiovascular risk factors, and four tested positive for a coagulation disorder. All patients received intravenous heparin. No obvious diffuse atherosclerosis or debris within the aortic arch on TEE examination was found. However, the thrombus insertion site involved an atheromatous plaque on the surgical specimen of 10 patients and on the autopsy of another. The authors postulated that aortic arch atherosclerosis constitutes a spectrum of disease ranging from pure atherosclerotic debris floating in the aorta, most prevalent in elderly patients, to nearly pure thrombus formation, often found in younger patients. Neither CT nor TEE can identify early atherosclerotic lesions, and the differential diagnosis of an isolated mass in the thoracic aorta should include vascular malignancies such as angiosarcoma. In summary, thoracic aortic thrombus should always be considered in young patients with unexplained stroke or peripheral embolism. Optimal management remains controversial due to a

Learning points

- ▶ Thoracic aortic thrombus should always be considered in young patients with unexplained stroke or peripheral embolism, and imaging of the thoracic aorta and great vessels of the neck, along with transesophageal echocardiography, is warranted.
- ▶ Anticoagulation therapy with intravenous heparin, followed by warfarin, is an effective first-line therapy.
- ▶ Anticoagulation duration in this setting remains unclear.
- ▶ Anticoagulation treatment can be given without delay, even in presence of large brain infarction. Careful follow-up and frequent brain CT or MRI must be performed to detect any developing intracranial bleeding.

paucity of experience. However, anticoagulation with intravenous heparin followed by warfarin has proved effective as a first-line therapy in many patients. The anticoagulation duration (until complete resolution or much longer) and target INR in patients on warfarin are unknown. The decision to proceed with surgery if thrombus persists depends on location and morphology of the thrombus, and the patient's overall clinical status. Recurrence rate after thrombus resolution is unknown and remains a major concern. Finally, the role of novel oral anticoagulants in this setting remains untested.

Competing interests None declared.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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