Mural aortic thrombus and peripheral embolisation in a patient with hyperhomocysteinaemia

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ABSTRACT

A mobile thrombus of the descending thoracic aorta in young people is extremely uncommon. We describe a 38-year-old woman with a mural thrombus in the proximal aorta complicated by peripheral embolisation, due to hyperhomocysteinaemia.

KEYWORDS

Aortic thrombus, embolisation, hyperhomocysteinaemia

INTRODUCTION

Homocysteine has been identified as an independent risk factor for atherosclerosis. It can lead to both venous and arterial thrombotic disease. We report on a young woman with hyperhomocysteinaemia and a mural thrombus of the proximal descending aorta without identifiable atherosclerotic lesions.

CASE REPORT

Two weeks after her third delivery, a 38-year-old woman came to the emergency room because of abdominal pain radiating to the shoulders of sudden onset. Her medical history was remarkable for a 20 pack-year smoking habit and a history of pregnancy complications.

Her first pregnancy at the age of 35 years was complicated by pulmonary embolism during the second trimester. At 33 weeks of pregnancy the foetus was lost due to a partial lissencephalia in combination with intracranial bleeding, as a result of high levels of anticoagulation. The second pregnancy was uncomplicated; she was then on low-molecular-weight heparin (LMWH).

In her third pregnancy, she stopped the LMWH therapy herself directly after delivery. She continued smoking during this pregnancy. One week after discontinuing the LMWH, the abdominal pain developed. Physical examination revealed normal blood pressure, pulse rate and temperature. Auscultation of lungs and heart was normal. Palpation of the left upper part of the abdomen was tender without palpable mass. All peripheral pulses were palpable. Striking laboratory results included a high sedimentation rate (102 mm/h) and a lactate dehydrogenase (LDH) of 900 U/l. Cholesterol was normal. A computed tomography (CT) scan did not reveal pulmonary embolism, but a low-density intraluminal lesion, 0.5 to 1.0 cm in size, was seen on the anterior wall of the proximal descending aorta (figure 1). Magnetic resonance imaging (MRI) confirmed these findings and revealed a large splenic infarction and a smaller renal infarction on the left side (figure 2). Transoesophageal echocardiography also showed a highly mobile mass, 0.5 to 1 cm in size, in the very proximal descending aorta. There were no signs of intracardiac thrombi, valvular disease, dilatation of the ventricles or abnormal systolic function. In view of these findings the patient was admitted to hospital for intravenous anticoagulant therapy.

Search for a hypercoagulable state revealed normal thrombin, prothrombin and partial thromboplastin times, as well as antithrombin III, protein C and S antigens, protein C pathway, and activated protein C resistance. Screening for anticardiolipin antibodies
and lupus anticoagulant was negative. Factor VIII was markedly elevated (405%) as frequently seen in the course of pregnancy. Postload plasma homocysteine level was elevated (68 umol/l, normal <45 umol/l). Vitamin B12 and folic acid levels were normal. A total body gallium SPECT scan was performed and showed no abnormalities. Media and intima thickness of the carotid arteries, measured by ultrasound, were normal.

In view of the hyperhomocysteinaemia, treatment was started with folic acid (0.5 mg once daily) and pyridoxine (20 mg once daily). The sedimentation rate and LDH normalised. She was discharged from hospital after two weeks. After two months transoesophageal echocardiography showed resolution of the thrombus and the nonfasting homocysteine blood level normalised (7 umol/l, normal <12 umol/l).

**DISCUSSION**

This young patient presented with acute onset of abdominal pain radiating to the shoulder. Taking into account her prior pulmonary embolism, the first differential diagnosis was a new pulmonary artery thrombus after she had stopped anticoagulant treatment. The CT scan in combination with the MRI of the aorta revealed unusual findings: ischaemic regions in the spleen and left kidney probably caused by emboli of a small mural thrombus in the proximal descending aorta.

A mobile intraluminal thrombus of the descending thoracic aorta is an unusual cause of peripheral embolisation. In this patient aortitis or vasculitis was excluded by gallium SPECT scan and atherosclerosis was unlikely in the light of physical examination, normal media and intima thickness and absence of calcifications in the large arteries on the CT scan. Primary aortic tumours may present with peripheral emboli but are extremely uncommon. The diagnosis is most often made after surgery or autopsy while growth is aggressive. CT, MRI and ultrasound did not show any solid masses except the thrombus. Although rare, mural aortic thrombi must be considered in a patient with otherwise unexplained peripheral embolisation. Most patients with this phenomenon have several risk factors for atherosclerosis. A high blood level of homocysteine has been identified as a risk factor for venous and arterial thrombotic disease. Patients with a mild hyperhomocysteinaemia are at risk of atherosclerotic vascular disease independent of diabetes, smoking, hypertension and hyperlipidaemia. The association with arterial thrombosis in the absence of atherosclerosis has rarely been described. It has been suggested that homocysteine can stimulate thrombosis by enhanced tissue factor expression and factor V activity in combination of suppression of thrombomodulin activity and decreased fibrinolysis. Furthermore, increased oxidative damage and proliferation of vascular smooth muscle cells have been noticed. The mobile aortic thrombus in our young patient without atherosclerosis...
strongly supports the diagnosis of a hypercoagulable state. A high homocysteine blood level was the only prothrombotic abnormality besides her pregnancy. Nevertheless our patient is a smoker. Graham described a two- to four-fold increased risk of cardiovascular events for smokers in the presence of hyperhomocysteinemia.

In this case, smoking could certainly have promoted the hypercoagulable state. Anticoagulation is an effective treatment for aortic mural thrombi. In this case the thrombus was completely resolved within two months. To the best of our knowledge, only one other case report has mentioned a patient with hyperhomocysteinemia, aortic thrombus and peripheral embolisation. That patient died before institution of treatment. With this case we provide clinical evidence to support the relationship of hyperhomocysteinemia and mural thrombus in the proximal aorta even without signs of atherosclerosis. Hyperhomocysteinemia should be considered when evaluating peripheral arterial thrombosis in a young person especially when atherosclerosis is absent.

ACKNOWLEDGEMENT

The authors thank Dr A.P.C. van de Maas for his critical comments on the manuscript.

REFERENCES