A Gangrenous Toe!

INTRODUCTION

This case is based on a patient who was found to have coincidental Mönckeberg’s disease when their peripheral arterial disease was being investigated. This should be something to look out for, since despite being asymptomatic, Mönckeberg’s is associated with increased arterial stiffness and increased pulse pressure, exaggerating damage to the heart and kidneys.

CASE REPORT

History

A 76 year old gentleman presented to the outpatient clinic with a 6 month history of left fifth toe gangrene associated with bilateral shooting rest pain in his feet. The pain was worse on movement and had been worsening in severity for one week. The patient also has multiple co-morbidities including coronary artery disease, known peripheral arterial disease, stroke, hyperlipidaemia and atrial fibrillation. He underwent a right renal transplant in 2003 and has a cardiac stent. He is a non-smoker and non-alcohol drinker. The patient has no known drug allergies and is taking several medications for relevant medical problems.

Examination

On physical examination, both feet appeared hyperaemic. There was no noticeable hair loss but there was a small, superficial ulcer on the left first toe, a small eschar on the right fifth toe and a dry gangrene on the left fifth toe. (Figures 1, 2 & 3) The peripheral pulses were weak, capillary refill time was less than two seconds and bilateral femoral pulses were present. The pulse was +1 in the dorsalis pedis and posterior tibialis on the right. The pulse was non-dopplable in the left dorsalis pedis but present in the posterior tibial artery. There was bilateral lower extremity oedema up to the mid-calf on the left and up and to the foot on the right of +2. Sensation was present bilaterally in both lower limb extremities. Initially, peripheral arterial disease (PAD) was suspected.

Investigations & Management

A computed tomographic (CT) angiogram without bilateral aortic run off showed the left common femoral artery, superficial femoral artery, profunda femoral artery, popliteal arteries, anterior tibial artery, posterior tibial artery and peroneal arteries were patent without stenosis or dissection. The trifurcation vessels were also patent. The dorsalis pedis arteries were diminutive and there was paucity of small vessels in the foot on imaging, likely secondary to peripheral arterial disease, causing the ulcer in the left fifth toe. Atheroembolic disease and calcification was noted in the common femoral artery and superficial femoral artery secondary to Mönckeberg’s disease, which was incidentally found. (Figures 4, 5, 6 & 7)

Lower extremity ultrasound was done using a colour doppler on the right which showed triphasic waveforms in all arteries except the dorsalis pedis, which displayed a monophasic waveform. Diffuse areas of calcific plaque was present in the right superficial femoral artery but the stenosis was not haemodynamically significant.

This patient was admitted. He was stable and apyrexial. He had a Body Mass Index (BMI) of 21.32kg/m². Blood results revealed microcytic anaemia with a haemoglobin of 9.2g/dL and mean cell volume of 75.4fl. He also had a low white cell count of 5.1 x 10⁹/L. The prothrombin time was raised at 16, which was likely to be due to warfarin. The patient’s calcium was 8.6 mg/dL, just slightly below the normal range. He was normoglycaemic.

Five days later, the patient underwent a left 5th toe amputation by vascular surgery and continued 75mg clopidogrel for peripheral arterial disease.

Figure 1

DISCUSSION

In Mönckeberg’s medial sclerosis (MMS), the diameter of the lumen remains preserved as the tunica intimia is largely unaffected.[1]

The exact molecular mechanism of vascular calcification remains unknown but the molecular pathway involved in vascular intima calcification differs to media calcification.

Persistent DNA damage signalling associated with cellular senescence can activate osteogenic pathways in vascular smooth muscle cells, making ageing the most dominant risk factor for the progression of tunica media calcification.

MMS has several other risk factors that may have contributed to this case including end-stage renal disease and type 2 diabetes mellitus. This patient had a renal transplant in 2003 and prior to this he had undergone haemodialysis. A rise in glucose, uremic toxins, phosphate and a drop in calcification inhibitors is believed to contribute to calcification.

Other risk factors for MMS are metabolic calcium disorders, congenital conditions, coagulation disorders and autoimmune disease.

Current treatment involves targeting the underlying causes such as diabetes mellitus, renal disease, abnormal metabolic bone disorders.

Age cannot be treated. Calcification increases exponentially with age and over two thirds of patients over 70, have tunica media calcification.

In advanced stages of MMS, there are alterations in autoregulation of blood pressure: increased pulse wave velocity, pulse wave pressure and pulse wave deformations.[6]

The ankle brachial pressure index (ABPI) is the most important screening tool to identify the presence of both peripheral arterial disease (PAD) and MMS.

However in this case ABPI was not necessary as RAD arteriogram was done. Rail tracking is seen in Mönckebergs typically due to media calcification. (Figures 8 & 9)[1]

MMS tends to occur in large elastic arteries with a diameter greater than 0.5 mm, rather than transitional or muscular arteries, coinciding with the fact that it occurred in the common femoral and superficial femoral artery in this patient.

Tunica media and intima calcification usually occur together, so many patients with MMS may also have peripheral arterial disease.

Compared to calcification in the tunica intimia, calcification in the tunica media is linear, always consists of calcium and tends to be non-inflammatory.[2]

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References: