



A Non-Atherogenic Cause of Critical Limb Ischemia in a Young Adult

**Jagadeesh K. Kalavakunta^{1,2*}, Yashwant Agrawal², Elizabeth Sayers³,
Fadi Saab³ and J. A. Mustapha³**

¹*Department of Cardiology, Borgess Medical Center, Kalamazoo, MI, USA.*

²*Department of Medicine/Pediatrics, Western Michigan University Homer Stryker M.D. School of
Medicine, Kalamazoo, MI, USA.*

³*Department of Internal Medicine/Cardiology, Metro Health- University of Michigan Health, Wyoming,
MI, USA.*

Authors' contributions

This work was carried out in collaboration between all authors. All authors read and approved the final manuscript.

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Case Study

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ABSTRACT

Aim: Atherosclerosis is the most common etiology of obstruction of the arteries leading to limb ischemia. Critical limb ischemia (CLI) in a young adult we need to consider non-atherosclerotic diseases.

Case Presentation: We present a rare of cause CLI in a 35-year-old man who presented with Rutherford class VI symptoms of non-healing gangrenous right great toe. His angiograms revealed medial type of fibromuscular dysplasia (FMD) and underwent endovascular therapy. Multiple interventions with balloon angioplasty, laser atherectomy and Ultrasound-accelerated catheter-directed thrombolysis in three different settings were needed to achieve desired results.

Discussion and Conclusion: In most cases of FMD balloon angioplasty itself is sufficient. But below knee arteries might need more than one intervention as noted in this case.

Keywords: Fibromuscular dysplasia; lower extremity; young adult.

1. AIM

Atherosclerosis is the most common etiology of obstruction of the arteries leading to limb ischemia. When critical limb ischemia (CLI) in a young adult presents, non-atherosclerotic diseases must be considered.

2. CASE PRESENTATION

A young Caucasian man in his mid-thirties was referred to the vascular clinic with complaints of non-healing gangrene of the right foot. He noticed discoloration of his great toe and thought that he might have bruised or may be due to exposure to cold as he lives in a cold climate. But it progressed in two weeks to gangrenous appearance, which prompted him to go to his primary care physician. He denied any trauma or prolonged exposure to cold. He had a history of <2 pack years of smoking. Past medical, surgical and family history was not significant.

Physical examination revealed a gangrenous appearance of the right foot indicating Rutherford class VI peripheral arterial disease (Fig. 1.). Pulses were 3+ on the left lower extremity. Right dorsalis pedis pulse was absent, posterior tibial was barely audible with hand held Doppler and the rest of the lower leg pulses were 2+. He was admitted to the hospital urgently and placed on a heparin drip. Duplex ultrasound of the right lower extremity showed triphasic wave forms external iliac (EIA), common femoral (CFA), superficial femoral (SFA), popliteal (POP), tibioperoneal trunk (TPT) with velocities (cm/s) 147, 178, 137, 89, 96 respectively and monophasic waveforms posterior tibial (PT) and peroneal artery (PA) with reduced velocities (cm/s). Anterior tibial (AT) and Dorsalis pedis (DP) had no flow.

Endovascular therapy was initiated after explaining risk, benefits and alternatives. Based on ultrasound results dual antegrade and retrograde access was deemed appropriate. Utilizing ultrasound a pedal dilator was placed into the right posterior tibial artery (PT) followed with a V18 wire (Boston Scientific). Access was obtained in the right common femoral artery (CFA) under ultrasound guidance and a 5 French (Fr) sheath was placed. Heparin was used for anticoagulation and activated clotting time (ACT) was maintained >200 throughout the procedure. Angiograms were obtained through the 5Fr sheath and a NaviCross (Terumo) catheter was

then advanced into the distal SFA. Selective angiograms were obtained which confirmed chronic total occlusions (CTOs) of the peroneal, PT and anterior tibial arteries (AT). Popliteal artery and peroneal artery showed a string of beads appearance suggestive of fibromuscular dysplasia (FMD) (Fig. 2.). The V18 wire from the right PT was used to cross the PT chronic total occlusion and tunneled into the antegrade NaviCross catheter and externalized. The pedal dilator was then removed. A 0.014" Regalia XS 1.0 wire (ASAHI) was placed into the right CFA to the level of the PT. In a stepwise fashion, multiple balloon dilatations were used to treat from popliteal to the lateral plantar arteries. A 7 Fr. Pinnacle destination (Terumo) sheath was placed into the right CFA. A 0.014" Command wire (Abbott Vascular) was advanced to the level of the AT. A Regalia wire was also placed and advanced to the level of the PT. Excimer laser atherectomy was then used to treat the tibioperoneal trunk (TPT), PT and lateral plantar arteries. This was then followed with 3.0 mm x 300 mm balloon angioplasty from the proximal PT to the lateral plantar. Direction was then given to the AT. A CXI catheter (Cook Medical) was placed over the Command wire. Laser atherectomy was used to treat the AT into the dorsalis pedis artery (DP). Multiple balloon dilations were performed in a stepwise fashion to treat the pedal loop, DP and AT. Repeat angiography revealed significant residual stenosis and spasm. The PT and lateral plantar arteries were treated again with a 2.0 mm x 220 mm balloon secondary to spasm with improvement in flow to TIMI III. Laser was then used to treat the AT and DP again followed by multiple balloon dilatations. Then Regalia wire was redirected into the peroneal artery. Laser atherectomy was used to treat the peroneal artery followed by percutaneous transluminal angioplasty (PTA). Repeat angiography revealed residual spasm, which was treated with nitroglycerin. Overall vessel stenosis was not reduced secondary to severe spasm. The 7 Fr Destination sheath was changed out to a short 7 Fr sheath which was to be removed when the ACT was deemed appropriate. The patient was brought back to the cath lab the next day and angiograms were repeated. A lot of thrombus burden in the peroneal, PT and AT was noted, hence an ultrasound-accelerated catheter-directed thrombolysis (Ekos, Ekos Corporation, Bothell, WA) initiated for 24 hours. Pedal pulses improved and pain was controlled well with

analgesics. A right foot x-ray was performed which did not show osteomyelitis but soft tissue swelling was noted in the mid and forefoot especially medial to the proximal phalanx of the great toe. 24 hours later repeat angiogram of the right leg was performed. Both AT and peroneal arteries were widely open but the PT was occluded which recoiled in spite of repeat balloon angioplasty. As the patient was doing well and the AT was open, which supplies the great toe region, the operators opted to treat conservatively. He was discharged from the hospital and has been doing well at his regular out-patient follow up visits.

3. DISCUSSION

In young patients who present with critical limb ischemia (CLI) non-atherosclerotic causes such as FMD, thromboangiitis obliterans (TAO), cystic adventitial disease of the popliteal artery, popliteal artery entrapment syndrome, iliac artery endofibrosis and vasculitis should be suspected. Here we discuss FMD in detail.

FMD is a rare non-atherosclerotic, non-inflammatory and multifocal arteriopathy affecting medium and large caliber vessels. It has been more than 75 years since its first description; however, the exact etiology is not known [1,2]. It commonly affects the renal and cephalic arteries but involvement of lower extremities is extremely rare. External iliac arteries are the most commonly involved arteries in the lower extremities with femoral, popliteal, AT, PT and peroneal arteries involvement extremely rare. Less than 15 FMD case reports involving the lower extremity arteries have been reported in the literature to the best of our knowledge.

FMD has been classified into three types depending on which arterial layer is involved-intimal, medial and adventitial type [3]. Medial type of FMD is the most common with string-of-beads appearance on the angiography. The beaded appearance is due to focal areas of stenosis due to fibromuscular ridges with intervening areas of aneurysmal dilatation. The classic angiographic appearance is only visible at the early stage of the disease. Once the vessel is occluded, inflammatory and atherosclerotic features makes FMD diagnosis difficult. In our case, we noted the string of beads appearance of medial FMD in popliteal and peroneal arteries. With the clinical presentation and angiography medial type of FMD was diagnosed in the present case.

The exact incidence of FMD involving the lower extremities is not clear. It predominantly affects women. FMD natural history is usually benign except in few patients. Genetic, hormonal, mechanical, smoking and mural ischemia have been proposed in the pathogenesis. Familial FMD is noted <10% in US registry for FMD [4]. Lower extremity FMD clinical presentation ranges from no symptoms to disabling symptoms such as claudication, limb-threatening ischemia, microembolic features or aneurysm rupture. FMD causes symptoms through stenosis, distal embolism or thrombus formation.

Diagnosis is made by clinical and imaging findings. It can be identified with Duplex ultrasonography, magnetic resonance and computerized tomographic arteriography but angiography is the standard for the diagnosis. In the angiography it can present as classic string-of-beads, focal stenosis or aneurysms. Before confirming the FMD by angiography we need to rule out arterial standing waves by giving vasodilators [5]. In our case, we did administer multiple aliquots of nitroglycerin in spite of that classic string-of-beads appearance persisted suggestive of FMD.



Fig. 1. Right foot showing the gangrenous great toe

Treatment depends on the symptoms and extent of arterial involvement. Asymptomatic patients are managed conservatively. Symptomatic patients have been treated with endovascular therapy with balloon angioplasty, thrombolytic therapy, anticoagulation, surgical options such as bypass surgery, aneurysm resection and rarely lumbar sympathectomy [6]. In the BASIL study, surgical bypass and balloon angioplasty have shown similar outcomes in terms of amputation and survival rate in 5.5 years follow-up [7]. There are no specific guidelines for treatment of lower extremity FMD due to lack of high quality data



Fig. 2. Angiogram of the popliteal artery showing the classic “string-of-beads” appearance before and after nitroglycerin

and low prevalence of the disease. Currently with the experience in renal arteries and external iliac arteries, percutaneous balloon angioplasty is considered as the first line therapy [8].

4. CONCLUSION

Fibromuscular dysplasia involving below knee arteries might need more than one intervention as noted in this case.

CONSENT

As per our hospital policy, since no identifiers/PHI are included, patient written informed consent is not required.

ETHICAL APPROVAL

As per our hospital policy, since this case study is not human subject's research, IRB review and approval is not required.

COMPETING INTERESTS

Authors JKK, YA, ES have declared that no competing interests exist pertinent to this report. Authors FS and JAM have consulting agreements with Boston Scientific, Terumo, and Cook Medical.

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