Conflicts of interest
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Sandra E. Sepúlveda[1], María J. Medina[1], Jorge O. Barretta[2], Horacio A. Capelli[3]
Hospital de Pediatría Prof. Juan P. Garrahan.
Buenos Aires, Argentina.
[1] Department of Cardiology
[2] Department of Cardiac Surgery
[3] Chief of Cardiology

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Blue Toe Syndrome as Expression of Severe Atherosclerosis

Aortic atherosclerotic plaques are a source of embolic phenomena that can occur with cerebral, splanchnic, or peripheral manifestations.

Cholesterol embolization syndrome (CES) or atheroembolism is the less common manifestation of embolism of an atherosclerotic plaque, and its prevalence is underestimated. It may occur spontaneously or after an arterial endovascular procedure. Cholesterol embolization syndrome is classified as definite and possible. Definite CES presents with cutaneous signs such as livedo reticularis, blue toe syndrome (BTS), and digital gangrene with or without renal involvement. Possible CES only shows renal involvement, that is, serum creatinine >1.3 mg/dl, two weeks after catheterization with normal renal function before the procedure, without cutaneous lesions. Atheroembolism, usually of the abdominal aorta, includes fragments of atherosclerotic plaque that contain cholesterol crystals and fibrin and platelet thrombi leading to disseminated microembolism. Microembolism causes inflammation associated to mechanical occlusion, and both cause ischemia and necrosis. (1)

Blue toe syndrome is a dermatological manifestation of CES with a frequency between 35% and 96%, characterized by tissue ischemia secondary to atheroembolism causing occlusion of the small vessels in the extremities. It presents with focal areas of painful cyanosis in the extremities, surrounded by normal tissue perfusion and preservation of distal pulses. Embolism typically originates from an ulcerated atherosclerotic plaque or from aneurysms located in the aortoiliacfemoral system. It can occur spontaneously or due to several causes (endovascular procedures, vascular surgery, anticoagulation, fibrinolysis). It is important to make differential diagnosis with Raynaud's syndrome, lesions due to hypothermia and idiopathic digital arterial thrombosis, as well as establishing the presence of atheroembolism in BTS, because it is a recurrent phenomenon and can lead to limb amputation or death if the embolism is very extensive. (2)

Diagnosis is predominantly clinical, but definitive diagnosis is made with biopsy of muscle or skin showing cholesterol crystals. Laboratory findings are nonspecific, but reveal a strong correlation with eosinophilia. (1)

Regarding diagnostic imaging, various methods can be used. Doppler ultrasound detects aneurysms or plaques proximal to the affected vascular bed, determining the embolic source. Computed tomography angiography and magnetic resonance angiography reveal the cause and severity of the underlying lesion. Diagnostic arteriography can determine the cause of thrombosis, provide information about the proximal circulation, and accurate details of the extension of collateral circulation and distal flow to the areas occluded by the embolism. (2)

We report a case of definite CES with BTS as dermatological manifestation, in a 57-year-old male patient, with cardiovascular risk factors –hypertension, non-insulin-dependent diabetes mellitus, dyslipidemia and smoking–, and unremarkable past medical history, who was admitted to our center due to 12-hour history of painful cyanosis in the right foot toes (Figure 1 A). The physical examination confirmed lack of pedal pulse and reduced posterior tibial pulse in the right lower limb. Lab tests revealed leukocytosis with eosinophilia and 0.75 mg/dl creatinine. The electrocardiogram was normal.

Diagnostic arteriography showed an ulcerated plaque causing severe stenosis at the level of the distal abdominal aorta, involving the origin of the inferior mesenteric artery (Figure 1 B), severe stenosis in the origin of the right primitive iliac artery, two severe stenoses in the origin and distal portion of the left primitive iliac artery (Figure 1 C), and opacification of the posterior tibial artery in the right foot; the anterior tibial and peroneal arteries were not visualized (Figure 1 D).

The condition was interpreted as BTS due to spontaneous atheroembolism as a result of ulcerated atherosclerotic plaque in the distal abdominal aorta, worsened by ischemia of the lower limbs secondary to severe stenoses described in both primitive iliac arteries.

It was decided to continue with endovascular treatment of the aorto-iliac axis disease.

Our case showed a large, ulcerated plaque in the distal abdominal artery as source of embolism. One of the options for endovascular resolution was covered stent placement to prevent embolization during the procedure. Covered stent implantation in the aorta was ruled out because the inferior mesenteric artery originated in the described plaque and would be occluded during stent placement. Bare self-expanding nitinol stents were chosen instead.

Access was performed with an 8F introducer in the right femoral artery and a 6F introducer in the left femoral artery, and the described stenoses were managed with bilateral 0.035" hydrophilic guidewire. A 14 mm wide by 60 mm long stent in the distal aorta, a 9 mm wide by 60 mm long stent in the right primitive iliac artery, and two stents, one 9 mm wide by 40 mm long and the other 8 mm wide by 40 mm long, in the left primitive iliac artery, were implanted. The aortic stent was dilated with a 10 mm balloon and the procedure was finished with a kissing balloon at the level of the aorto-iliac bifurcation with two 8 mm balloons, with positive angiographic outcome and without involving the origin of the inferior mesenteric artery (Figure 2).

The patient evolved with no toe pain, persistence of cyanosis, recovery of pedal pulse, and marked improvement of posterior tibial pulse. The patient was discharged 72 hours after the procedure with dual antiplatelet therapy, statins, vasodilators, and hypoglycemic agents.

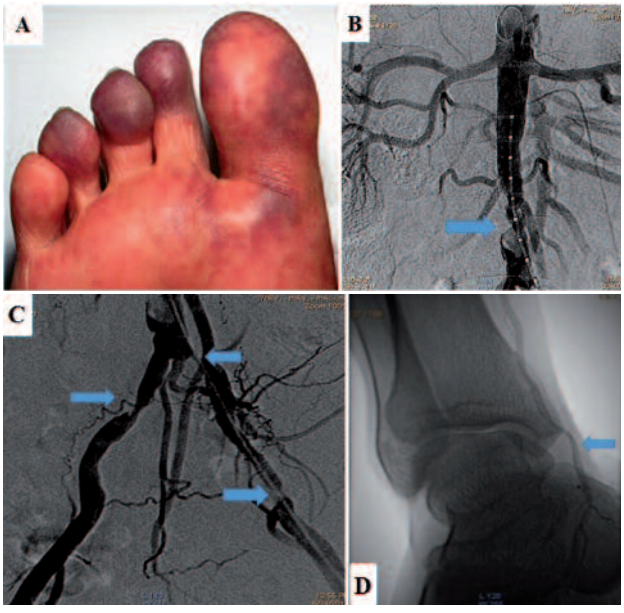

Fig. 1. A. Right foot toes showing cyanosis. **B.** Ulcerated atherosclerotic plaque in distal abdominal aorta involving the origin of the inferior mesenteric artery (*arrow*). **C.** Severe stenosis in both primitive iliac arteries (*arrows*). **D.** Distal circulation. Only the right posterior tibial artery shows opacification (*arrow*).

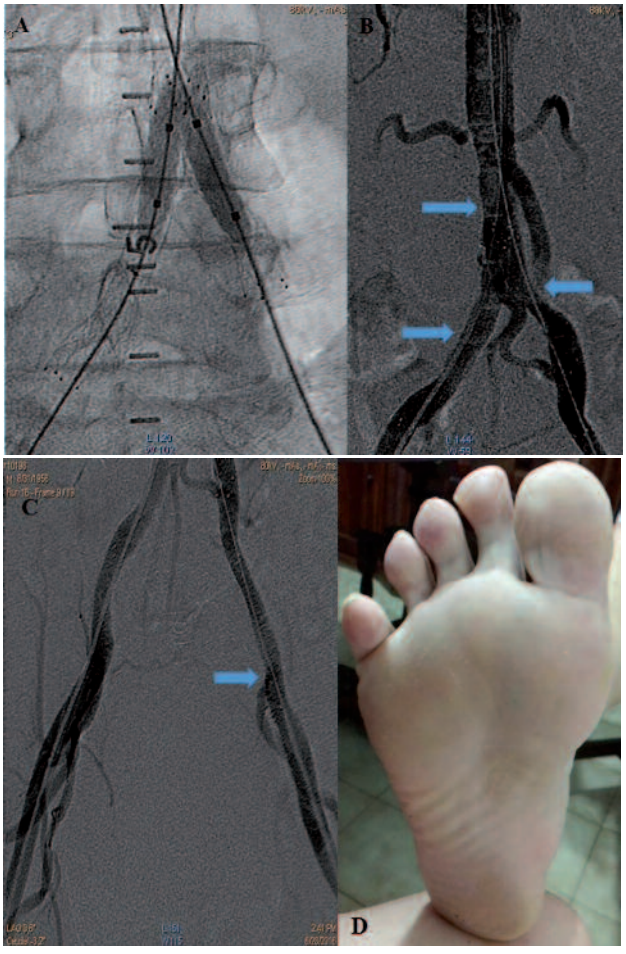

Fig. 2. A. Kissing balloon placement is observed. **B & C.** Angiography shows implanted stents and absence of inferior mesenteric artery involvement (*arrows*). **D.** Complete resolution of cyanosis.

At 10-month follow-up, the patient has made good progress with no recurrent embolic phenomena, complete resolution of cyanosis in right toes (Figure 2 D), preserved distal pulses, and arterial Doppler ultrasound showing triphasic flow in the right lower limb.

There is no specific therapy for BTS. Supportive measures to prevent the progression of atherosclerotic disease include modifications of risk factors, use of statins and antiplatelet agents, and avoidance of anticoagulation. (3)

Traditionally, the treatment options for BTS include endarterectomy or bypass to exclude the embolic source. Due to the bad prognosis of patients with BTS, there is still much controversy about the best treatment for these patients. For this reason, various studies focusing on the resolution of the entity have been published. Stents proved to be effective in the treatment of this arterial condition, providing a platform to prevent future embolisms and promote atherosclerotic plaque remodeling. Although there is concern for increasing distal embolization during stent placement, there is no clinical evidence to demonstrate it in the period immediately after placement of the device. (4-6)

Blue toe syndrome is a relatively rare condition, but it is a manifestation of atherosclerotic disease that can be devastating due to its morbidity and mortality. Cardiologists should bear this entity in mind when encountering a patient with the clinical manifestations described above, since early revascularization can prevent limb amputation or patient death.

Conflicts of interest
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Marcel G. Voos Budal Arins, Jorge N. Wisner[MTSAC], Alejandro Tettamanzi, Esteban A. Mendaro, Laura V. De Candido
Interventional Cardiology and Hemodynamic Unit
Hospital Universitario CEMIC. Buenos Aires, Argentina.
Marcel G. Voos Budal Arins.
E-mail: marcelvoos@gmail.com

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