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Blue toe syndrome: A challenging diagnosis

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Full Text

Sir,

"Blue toe syndrome" (BTS) refers to the acute onset of purple painful digits in the absence of evident trauma, cold-associated injury or disorders that induce generalized cyanosis. The term was used for the first time in 1976 by Komody, who underlined the vascular etiology of the disease and its possible diagnostic confirmation through angiography.[1],[2] Indeed, BTS may occur from end-arterial occlusion, impaired venous outflow, and/or abnormal blood circulation. Peripheral microembolism with distal arterial occlusion is one of the most frequent underlying mechanisms of the disease and consists of disrupted material from ulcerated atheromatous plaques (atheromatous or cholesterol crystal emboli). The case described by us assumes significance because of an atypical clinical presentation of a peripheral embolism from an abdominal aortic aneurysm, hence necessitating a high index of suspicion to achieve the correct diagnosis.

An 82-year-old man showed up with a painful chronic ulcer located on the second toe of his left foot. The lesion had been present for 3 months and was worsening. He had hypercholesterolemia, hypertension, and a personal history of trauma during World War II, with bomb fragments wedged inside his right foot. Previous noninvasive vascular assessment showed an abdominal aortic aneurysm, stable at a previous ecographic follow-up, while last year, a peripheral ecographic examination was normal. Upon physical examination, a small wound was observed on the second toe of the left foot [Figure 1]. Even though peripheral pulses were intact, all toes were cold, painful at palpation, and showed a cyanotic color and livedo reticularis. Moreover, a clear abdominal pulse was detectable and palpable. A doppler exam was promptly performed and pointed out a normal flow in both dorsalis pedis arteries and a very compromised flow in the digital arteries. Laboratory testing was carried out and revealed eosinophilia (eosinophil count 1.05 × 109/L), anemia (Hb 10.1 g/dL),

and a declined renal function (serum creatinine concentration 1.84 mg/dL). The patient was referred to a radiology center for an abdominal computed tomography angiography, in order to exclude a blue toe syndrome caused by peripheral embolism from the abdominal aortic aneurysm. The Computed Tomography (CT) scan showed a voluminous and partially thrombosed aortic aneurysm, with rehash of the thrombotic component. He was hospitalized in our Surgery Department. In a few days, serum creatinine concentration increased to 5 mg/dL and he was put on dialytic therapy. Surgical or endovascular treatment was not possible due to his advanced age and comorbidities. Two weeks later, serum creatinine concentration was improved; he was discharged with medical treatment.{Figure 1}

In BTS, physical examination can reveal livedo reticularis, gangrene, cyanosis, skin ulceration, purpura, and petechiae. Since embolism affects vessels of small diameter, peripheral pulses often remain detectable. Therefore, the simultaneous presence of distal gangrene/ulcers and palpable pulses are considered highly suggestive of blue toe syndrome. However, even if a presumptive diagnosis of cholesterol crystal syndrome may result from relevant clinical features, only skin biopsy with histological examination can definitively lead to a diagnosis of certainty.[2],[3],[4],[5] Embolization can be spontaneous or triggered by coexistent coagulation disorders, neoplastic processes, anticoagulant/fibrinolytic therapy, and/or invasive percutaneous/endovascular procedures.[4],[5],[6],[7] If the origin of the microemboli is proximal to the bifurcation of the abdominal aorta, cutaneous involvement tends to be early and bilateral, with cyanosis and livedo reticularis affecting several digits. According to the site of origin, emboli may also affect other body regions, such as kidneys, liver, spleen, pancreas, gastrointestinal tract, and adrenal glands, with mortality as high as 81% in cases of multiple organ involvement. Surgical or endovascular treatment, together with a correct management of cardiovascular risk factors, has been shown to be effective in reducing the rate of new episodes.[6]

In our case, the patient was studied for an aortic aneurism, stable in dimensions at follow-up. The anamnestic data of trauma and foreign body retention could lead to a wrong diagnosis. Doppler ultrasound confirmed our diagnostic suspicion, showing no pulse in most digital arteries while at the same time pulse and flow in the doralis pedis arteries.

Since BTS may be the first manifestation of multiple and potentially life-threatening conditions, its prompt recognition can avoid concerning consequences. Complete past medical history, physical examination, basic laboratory tests, and noninvasive vascular assessment must be carefully evaluated by dermatologists, since early cutaneous findings can play a fundamental role in initiating the correct BTS management.[7] However, a multidisciplinary approach is required to suggest the optimal medical or surgical therapeutic strategy.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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