Blue Toe Syndrome: A Peripheral Manifestation of Systemic Diseases

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INTRODUCTION:

Blue (purple) toe syndrome presents as a bluish discoloration of the toe due to occlusion of the small digital arteries by fibrino-platelet micro-emboli originating from a proximal atheromatous source cardiac, aortic, femoral or popliteal aneurysm. The peripheral pedal pulses are palpable excluding peripheral vascular disease. Embolization can occur spontaneously [2] or, following anticoagulant therapy rarely. When it occurs in patients with a recent history of an invasive vascular procedure or anticoagulant therapy, it is most likely due to cholesterol embolism. This article reports a case of Blue toe syndrome in this environment, reviews the aetiology and describes the clinical features. It also highlights the differential diagnosis of a blue toe.

CASE REPORT:

An 80 year old woman was noticed to have developed bluish discoloration of her toes 6 days after admission. She was being managed for acute confusional state in the elderly resulting from urosepsis. She was a known hypertensive diagnosed 15 years ago, with poor drug compliance. She was not diabetic and had been admitted repeatedly for heart failure. She did not smoke or take alcohol. Examination of the Cardiovascular system revealed tachycardia with an irregularly irregular pulse and the first heart sound had varying intensity. She was however not in heart failure. She was observed to have bluish discoloration of the tips of the second and fourth fingers on both sides and both halluces, she also had slightly cold extremities although both dorsalis pedis pulsation were palpable (Fig 1-3).

ECG done showed multifocal atrial tachycardia. Echocardiography showed dilated left atrium and ventricle with impaired systolic and diastolic functions, Ejection fraction (EF) was 39%, moderately severe mitral regurgitation, moderate tricuspid and pulmonary regurgitation and left ventricular spontaneous echo contrast - Echo smoke (Fig 4).

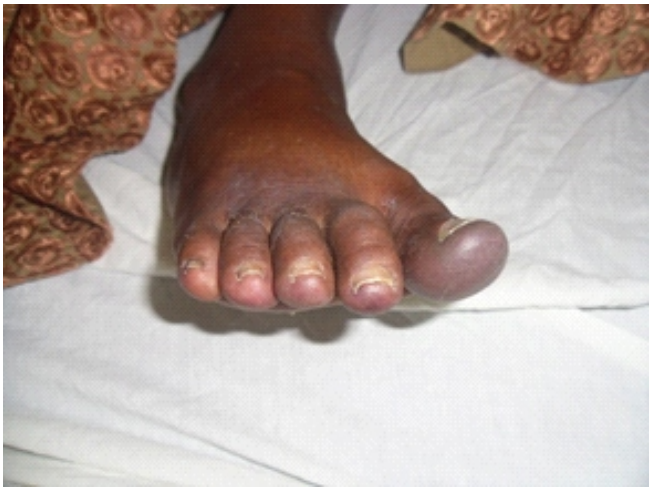

Figure 1

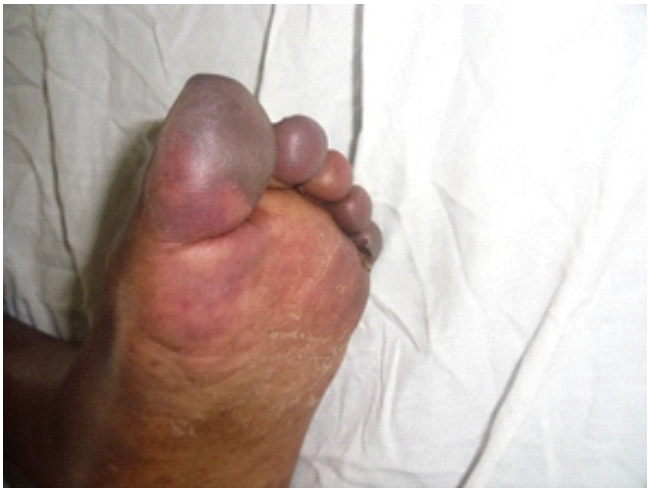

Figure 2

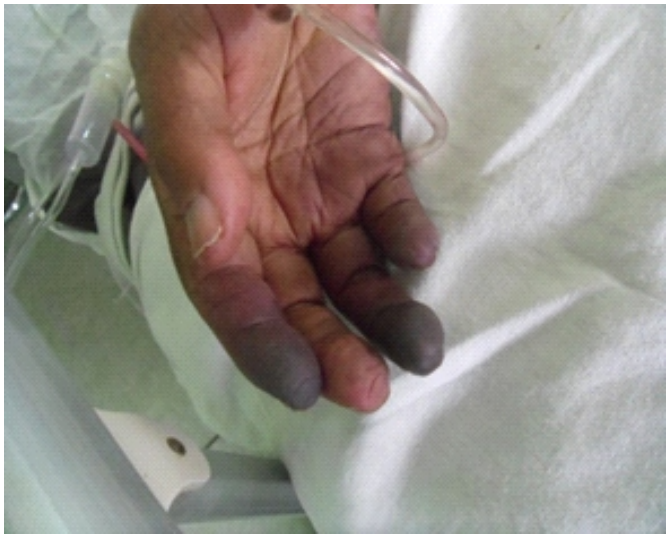

Figure 3: Blue fingers

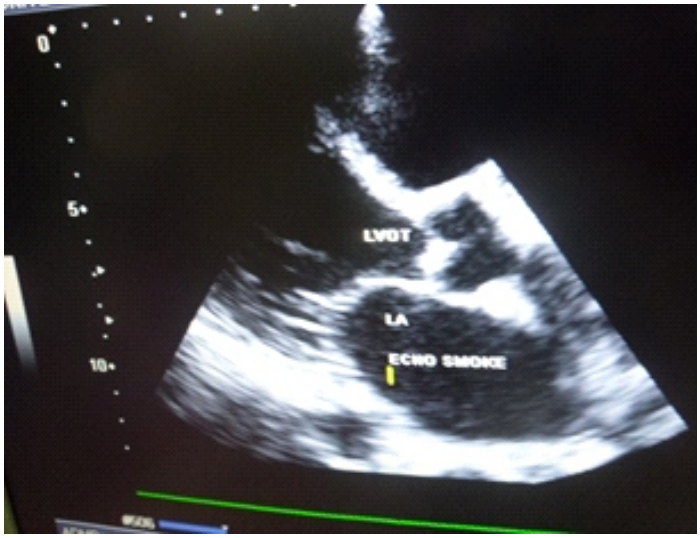

Figure 4: Echocardiography

Hand held Doppler examination showed that both dorsalis pedis were patent, biphasic and equal. Detailed Doppler examination was to follow.

She was reviewed by the Cardiology, CTSU, Haematology and Urology teams respectively and appropriate medications were prescribed by each team at the time of review. She however developed acute kidney injury and had a cardiac arrest on day 14 on admission.

Other investigations done: PCV: 41%, Electrolytes, Urea and Creatinine values were within normal values.

DISCUSSION:

Blue toe syndrome is the spontaneous onset of a painful bluish discoloration of the toes caused by fibrino-platelet micro-emboli from the abdominal aorto-iliac-femoral arterial system, which travels down the arterial tree into the small digital vessels of the foot, where it becomes lodged. The blue (or purple) toe syndrome develops in the absence of obvious trauma, serious cold-induced injury, or disorders producing generalized cyanosis.(2) The major disorders causing this micro-emboli formation can be grouped into three general categories: those due to decreased arterial flow, impaired venous outflow, and abnormal circulating blood. In blue toe syndrome skin lesions are usually restricted to the occluded artery. Clinical presentation can range from a cyanotic toe or livedo reticularis to a diffuse multiorgan systemic disease that can mimic other systemic illnesses. Acute focal digital ischemia may result from embolism, thrombosis, mechanical obstruction or inflammation or from a combination of these mechanism. Amongst other foci, is the intra-cardiac chamber thrombi can develop during the course of several cardiac pathologies that favor the reduction in blood flow as it occurs in acute myocardial infarction, left ventricular [LV] aneurysms, cardiomyopathies and myocarditis. Other causes include valve disease with or without prosthesis. Atrial fibrillation as seen in our patient may predispose to blood clots in the cardiac chambers. In a study by Vongpatanasin et al, it was documented that a potential cardiovascular source of embolism was the heart as demonstrated by transesophageal echocardiography in 20 of 33 patients (61%) with acute limb ischemia. The percentage was higher in patients with large artery occlusions (9 of 11, 82%) than in those with small artery occlusions (9 of 22, 41%) (p = 0.026). Patients with bilateral blue toe syndrome usually have an embologenic lesion proximal to an aortic bifurcation, while unilateral lesions are usually associated with an atherosclerotic plaque or aneurysm distal to the aortic bifurcation. A In patients with no obvious embolic source, systemic abnormalities should be considered, including circulating agglutinins, vasculitis, acrocyanosis, systemic lupus erythematosus, polycythemia vera and sub-acute bacterial endocarditis, as well as certain drugs such as warfarin and prednisolone. Untreated blue toe syndrome frequently recurs and could result in loss of a limb and death.

Echocardiography is important in the investigation of patients with AF as it diagnosis the presence of LA thrombus formation (up to 15% event/year among patients with AF). Our patient had dilated cardiac chamber, left ventricular hypertrophy and spontaneous echo contrast, defined as 'Echo smoke' on a transthoracic echocardiography (TTE). The

presence of spontaneous echo contrast, is the most evident sign of slackened blood flow. Spontaneous echo contrast is considered a prethrombotic condition, associated with an increased risk for thromboembolic events. A transesophageal echocardiography (TEE) could have been desirable to rule out possibilities of clots in the left atrial appendages (LAA) but was not available in our center. Notable is the fact that TEE is the gold standard tool as allows a good visualization of LAA and pulmonary veins, a complete exploration of atrial cavity

The therapeutic goal for blue toe syndrome consists of surgical or percutaneous elimination of the source of embolism. Medical treatment is mostly symptomatic: rest, warm condition, appropriate dressing, hydration, and organ support when necessary, particularly to ensure renal function. Treatment of pain that is usually disproportionate to the extension of tissue lesion is of utmost importance. Because these patients usually have advanced atherosclerotic disease, secondary prevention with elimination of risk factors of atherosclerosis is mandatory.

In summary, Blue toe syndrome may result from embolisation due to many clinical disorders, however early recognition will prevent loss of life and limb.

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