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Case Report

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Purple toe: a syndrome from head to toe

Abstract

Cholesterol crystal embolization is a rare multisystem disorder that originates from atherosclerotic plaques showering to different sites causing end organ damage. We present a 79-year-old man who developed purple discoloration of the left toe. Physical exam revealed tender purple toes with palpable distal pulses. The diagnosis of cholesterol emboli syndrome was made while differential diagnoses were ruled out. Despite medical management, his condition continued to slowly worsen.

Keywords: cholesterol emboli, purple toe syndrome

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Introduction

Cholesterol crystal embolization (CCE) is a rare multisystem disorder that originates from atherosclerotic plaques showering to different sites causing end organ damage. Strokes, specifically lacunar Infarcts, secondary to cholesterol cerebral emboli are extremely rare and barely reported in the literature. CCE is predominantly seen in elderly men (61%), and the most common findings in these patients are renal failure (50%) and cutaneous presentations (34%). Although other nonspecific findings like, fever, myalgia, headache or weight loss could be seen.[1]

Case presentation

A 79-year-old man, with history of HTN, CAD and dyslipidemia, presented with two-week history of painful toes that started as 'purple' discoloration on the left side then extended to involve most of his toes bilaterally. Two months prior to this presentation, he sustained a left internal capsule stroke along with concomitant worsening of kidney function. One month later, a change in the mental status was noted in the inpatient rehabilitation unit, and a CT head revealed a new left parietal stroke. Physical exam revealed purple toes with severe tenderness to touch and a dry small ulcer on the left forth toe (Figure 1).

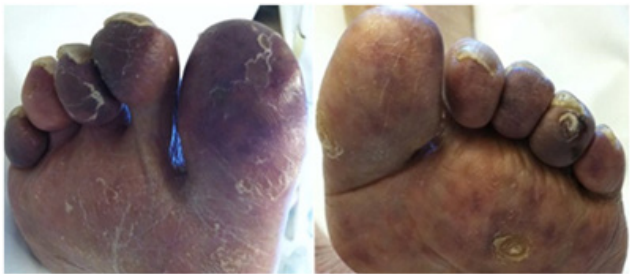

Figure 1 Purple discoloration of the bilateral toes of the patient with developing ulceration at the tip of the left toe resulting from underlying cholesterol embolization (Purple Toe Syndrome).

Dorsalis pedis pulses were detectable. Ankle-Brachial Index was 1.16 and 1.18 on the right and the left, respectively. Funduscopic examination showed bilateral cataracts. Laboratory workup showed eosinophilia and creatinine reached a plateau of 2.0 from a baseline of 1.3. Vasculitis workup including ANA, C-ANCA, P-ANCA, cryoglobulins and complements, was negative. A transesophageal echocardiogram revealed significant non-mobile atheromatous burden of the thoracic aorta and aortic arch. A computed tomographic scan of the abdomen and pelvis showed thoracic aortic aneurysm and infrarenal abdominal aortic aneurysm with 4.3 x 3.5cm dimensions. Clinical diagnosis of cholesterol emboli syndrome was made. The patient was managed with aspirin and statin. A trial of intravenous prostaglandin I2 for 48hours did not improve his symptom. Although the purple toes continued to slowly get worse, the patient survived and was discharged to a nursing home facility.

Discussion

Cholesterol crystal embolization is a rare multisystem disorder with immunologic features that originates from ulcerated atherosclerotic plaques, usually in the aorta, showering to different sites causing end organ damage.[2] The incident usually follows endovascular interventions but can occur spontaneously. In the latter setting, it could be an early sign of disseminated intravascular coagulation (DIC).[3] Classically, it presents with pain in legs before skin lesions become evident. Complications can be acute and catastrophic with multi-organ involvement including, bowel infarction, mucosal ulcerations and pancreatitis. Strokes, specifically lacunar infarcts, secondary to cholesterol cerebral emboli are extremely rare and barely reported in the literature. In our case, the acute infarct of the left parietal area was likely secondary to cholesterol emboli as its occurrence coincided with the acutely worsening renal failure and toe purplish discoloration. Histopathology remains the gold standard diagnostic method which can show cholesterol crystals within disrupted blood vessels appearing as arteriolar occlusion with needle-shaped clefts and surrounding foreign body giant cells. Eosinophilia and transiently low C3 and C4 complements may suggest the diagnosis but their absence should not rule it out. Other diseases such as vasculitis, endocarditis and cutaneous peri-arteritis nodosa, may need to be ruled out. Prognosis depends on complications but usually is self-limited. Treatment is generally medical and supportive including aspirin, statins, blood pressure and glucose control.[1] If there is inflammation and eosinophilia, steroid usage could be beneficial.[4,5] Prostaglandin I2 was associated with clinical improvement in some cases.[6,7] Anticoagulation should be avoided as it can lead to plaque instability and rupture with cholesterol emboli showering. Although purple toes was the presenting symptom leading to considering the diagnosis, it

indeed manifested with an acute ischemic stroke- a reason for which we consider this case extremely rare.

Conclusion

Cholesterol crystal embolization is a rare but underestimated syndrome. It is one of the "great imitators" and other diseases should be excluded. Stroke is an atypical presentation of this syndrome but should be considered beside the constellation of the other findings especially when there is lacunar infarction. Awareness of this syndrome can change the overall management and its ignorance may lead to unfavorable outcomes.

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Conflicts of interest

Author declares there are no conflicts of interest.

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