

# A 76-year-old male with a blue toe and livedo reticularis

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## CASE REPORT

A 76-year-old Caucasian male presented with acute kidney injury. His past medical history included hypertension (of unknown duration) and peripheral artery disease, for which percutaneous transluminal angioplasty of the right leg had been performed in 1997. He had smoked 38 pack-years. In the past three days he had experienced a painful right toe. He was not on any oral anticoagulants. On physical examination the patient was lean, not oedematous and his blood pressure was 120/69 mmHg. He had a marked cyanotic right toe (*figure 1*) with absent pedal pulses and livedo reticularis most prominently on the left knee (*figure 2*). Laboratory investigation showed an erythrocyte sedimentation rate of 44 mm/hour, mild leucocytosis of  $10.9 \times 10^9/l$  with an eosinophil count of

**Figure 2.** *Livedo reticularis* most prominently on left knee



**Figure 1.** *Cyanotic digit 3 of right foot*



$0.58 \times 10^9/l$ . Serum creatinine was  $213 \mu\text{mol/l}$  (baseline creatinine  $97 \mu\text{mol/l}$ ) with an estimated glomerular filtration rate of 26 ml/min. Urinalysis revealed 2.12 g/l of protein and no erythrocyturia. Antineutrophil cytoplasmic antibodies were negative. Ultrasound demonstrated relatively small kidneys of 9.1 cm and 9.5 cm and an infrarenal abdominal aneurysm. Computer tomography (CT) imaging showed no renal artery stenosis and confirmed the aneurysm, which measured 5.6 cm and contained atherosclerotic plaques and a mural thrombus.

## WHAT IS YOUR DIAGNOSIS?

See page 261 for the answer to this photo quiz.

## DIAGNOSIS

Based on the dermatological features (blue toe and livedo reticularis) and eosinophilia in addition to acute kidney failure, the diagnosis of spontaneous cholesterol crystal emboli (CCE) was made.<sup>1</sup>

CCE is iatrogenic in the majority of cases, with angioplasty (50%), vascular surgery (15%) and long-term anticoagulant therapy (76%) being the most common aetiological factors.<sup>2</sup> Spontaneous CCE are rare, with an estimated incidence of only 1.9%.<sup>3</sup> Typically, a patient with CCE is a lean, smoking male suffering from other manifestations of atherosclerosis.<sup>1</sup> Almost all patients with CCE (97%) have plaques in their thoracic aorta and 67% of patients have an aortic abdominal aneurysm.<sup>2</sup> Cholesterol emboli cause arterial occlusion leading to end-organ damage, involving most commonly the brain, kidneys, gastrointestinal tract, the skin and skeletal muscles of the lower extremities. Classic clinical features are livedo reticularis and cyanotic toes. Histopathology is regarded as the golden standard.<sup>4</sup> However, because on the one hand the combined clinical scenario and features in addition to the laboratory findings were very suggestive of the diagnosis of CCE and on the other hand performing a biopsy had no clear therapeutic consequences, a confirmatory kidney biopsy was not performed in our patient. Also, biopsy of the affected skin or skeletal muscles is performed infrequently because it may lead to poor healing at the sampling site.<sup>1</sup>

To date there is no specific therapy for CCE. Because this disorder is a manifestation of atherosclerosis, modification of traditional risk factors for atherosclerosis such as smoking, hypertension and serum cholesterol is strongly advised.<sup>1</sup> Angiotensin-converting enzyme inhibitors or direct angiotensin receptor blockers and antiplatelet therapy could be considered. Since anticoagulant therapy may aggravate CCE, these drugs are contraindicated.<sup>1</sup> Surgical therapy has been shown to be effective in decreasing the rate of future embolism. The role of corticosteroids in CCE is still inconclusive.

## REFERENCES

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