

Spontaneous Cholesterol Crystal Embolism – A Rare Cause of Renal Failure

Dear Editor,

A 68-year-old male chronic smoker with a history of long standing hypertension presented with a 3-month history of intermittent lower limb claudication, associated with bluish-purple discolouration of bilateral toes. He had no history of angiographic or vascular procedures, neither was he on anticoagulation treatment. On examination, his blood pressure reading was 125/65 mmHg. Bilateral blue toes were noted (Fig. 1) with well palpable dorsalis pedis. The remainder of the physical examination was unremarkable.



Fig. 1. Cutaneous manifestation of cholesterol crystal embolism (CCE) as blue toe syndrome.

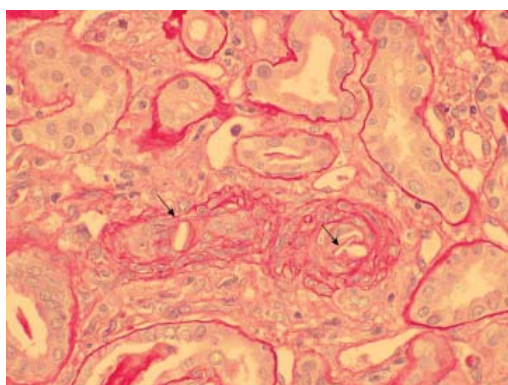


Fig. 2. Renal biopsy showed marked arteriolar nephrosclerosis, with classical intraluminal acicular transparent clefted spaces, consistent with cholesterol crystal embolism (CCE).

Laboratory investigations revealed an elevated serum creatinine of 1000 $\mu\text{mol/L}$. There was mild leukocytosis of 10,400/ mm^3 with eosinophilia (18%) and the haemoglobin concentration was 9.8g/dL. The C-reactive protein was 174 mg/L. Urinalysis was bland with trace proteinuria. Antineutrophil cytoplasmic antibodies and autoimmune markers were negative and complement levels were normal. Renal ultrasonography and doppler scan of the abdominal aorta showed increased renal echogenicity and diffused fibrocalcific plaques respectively. Renal biopsy showed cholesterol atheroembolism with moderate tubular atrophy (Fig. 2). Haemodialysis was initiated, however patient passed away within 6 months of diagnosis.

We describe a case of spontaneous cholesterol crystal embolism (CCE) as a cause of end-stage renal failure (ESRF). While CCE is iatrogenic in the majority of cases, with angioplasty or vascular surgery being the most common triggering events (reported as 50% and 15%, respectively),¹ spontaneous CCE is rare with estimated incidence of 2%.² It tends to affect male smokers in their sixties with underlying atherosclerosis, and manifests as a classic triad of livedo reticularis, renal failure and eosinophilia. Renal involvement is seen in half of the cases and gives rise to a spectrum of renal impairment, with the chronic subset often being underdiagnosed.³ Histopathological confirmation by biopsy remains the definite diagnostic test,³ however it is more recognised recently that the yield of non renal biopsies (skin, muscle, gastric and colon mucosa) is high and less invasive, and fundoscopic examination to look for retinal crystals may also provide a diagnostic opportunity.^{2,4} It follows that renal biopsy as the sole means of diagnosing CCE is only necessary in about 20% of cases.²

The clinical presentation when there is multisystem involvement often poses a diagnostic challenge. The spectrum involving renal, skin, gastrointestinal and central nervous system may mimic various types of vasculitis, with differential diagnoses including polymyositis and subacute bacterial endocarditis. To date, there is no curative treatment for this disorder. Therapeutic options are conservative and preventive, and prognosis remains guarded.^{2,3} Statin therapy may have a beneficial effect on outcome, but more trials are needed. The role of corticosteroid and antiplatelet agents in CCE is still inconclusive.^{3,4}

REFERENCES

1. Scolari F, Tardanico R, Zani R, Pola A, Viola BF, Movilli E, et al. Cholesterol crystal embolism: A recognizable cause of renal disease. *Am J Kidney Dis* 2000;36:1089-109.
2. Meyrier A. Cholesterol crystal embolism: diagnosis and treatment. *Kidney Int* 2006;69:1308-12.
3. Kronzon I, Saric M. Cholesterol embolization syndrome. *Circulation* 2010;122:631-41.
4. Scolari F, Ravani P, Gaggi R, Santostefano M, Rollino C, Stabellini N, et al. The challenge of diagnosing atheroembolic renal disease: clinical features and prognostic factors. *Circulation* 2007;116:298-304.

Kian Guan Lee,¹*MBBS (Malaysia), MRCP (UK)*, Hwai Liang Loh,²
MBChB(Hons), FRCPath, Chieh Suai Tan,¹*MBBS (Singapore), MRCP (UK)*

¹Department of Renal Medicine, Singapore General Hospital, Singapore

²Department of Pathology, Singapore General Hospital, Singapore

Address for Correspondence: Dr Kian Guan Lee, Department of Renal Medicine, Singapore General Hospital, Outram Road, Singapore 169608.
Email: lee.kian.guan@singhealth.com.sg