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Severe systemic calciphylaxis with culture-negative endocarditis

Tatsuhiko Azegami[1], Shu Wakino[1], Matsuhiko Hayashi[2] and Hiroshi Itoh[1]

[1]Department of Internal Medicine, School of Medicine, Keio University, Tokyo, Japan and [2]Apheresis and Dialysis Center, School of Medicine, Keio University, Tokyo, Japan

Correspondence and offprint requests to: Shu Wakino; E-mail: shuwakino@z8.keio.jp

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A 62-year-old male patient presented to the hospital with sustained fever and refractory skin ulcers. He had been under haemodialysis for 11 years and had received anticoagulation therapy via warfarin following the artificial valve replacement surgery for 4 years. From his painful skin ulcers and necrotizing skin nodules with surrounding purpura over a wide range of the inferior limb, perineum, penis and breech and penile gangrene (Figure 1), calciphylaxis was diagnosed by a criteria proposed by the Japanese Calciphylaxis Study Group [1]. On admission, the white blood cell counts were 19.0×10^9 /L and the C-reactive protein level was 160 mg/L. Skin culture revealed isolation of methicillin-resistant *Staphylococcus aureus*, while standard culturing of blood had never resulted in the isolation of any microorganisms. Administration of antibiotics was started and warfarin was replaced with continuous infusion of heparin, because warfarin is considered a risk factor for the aggravation of calciphylaxis [2]. Since prosthetic valves can be a focus for infectious endocarditis, transthoracic echocardiography was performed and verrucous vegetation on the mitral valve was detected (Figure 2a). Culture-negative infectious endocarditis was clinically diagnosed. Administration of 0.5 g of vancomycin and 80 mg of gentamicin was started. However, the infection was still uncontrolled and he died of septic shock 13 days after admission. Autopsy revealed a thrombus in the mitral valve with leukocyte infiltration consistent with the diagnosis of infectious endocarditis (Figure 2b). In the skin lesions, calcified arterial media with concentric intimal hypertrophy, the typical pathology of calciphylaxis, were detected throughout the body (Figure 3). Although infection is the main cause of morbidity and mortality for patients with calciphylaxis [3], superinfection in calciphylaxis with endocarditis is very rare. Only one case is reported in English-language literature [4]. This report stresses the need for exploring superinfection in the patient with calciphylaxis, including infectious endocarditis.

Conflict of interest statement. None declared.

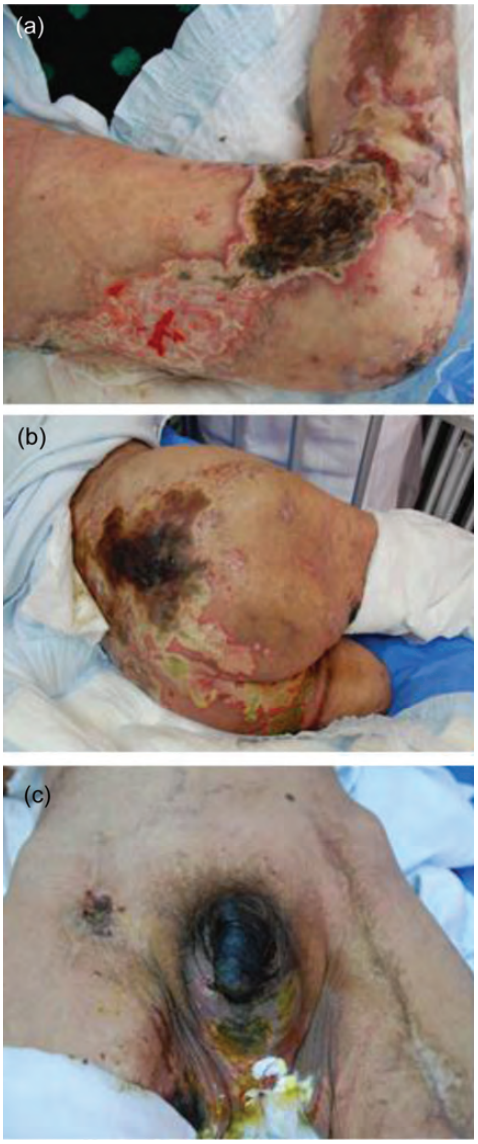

Fig. 1. Skin lesions of right inferior limb (**a**), breech (**b**) and penis (**c**).

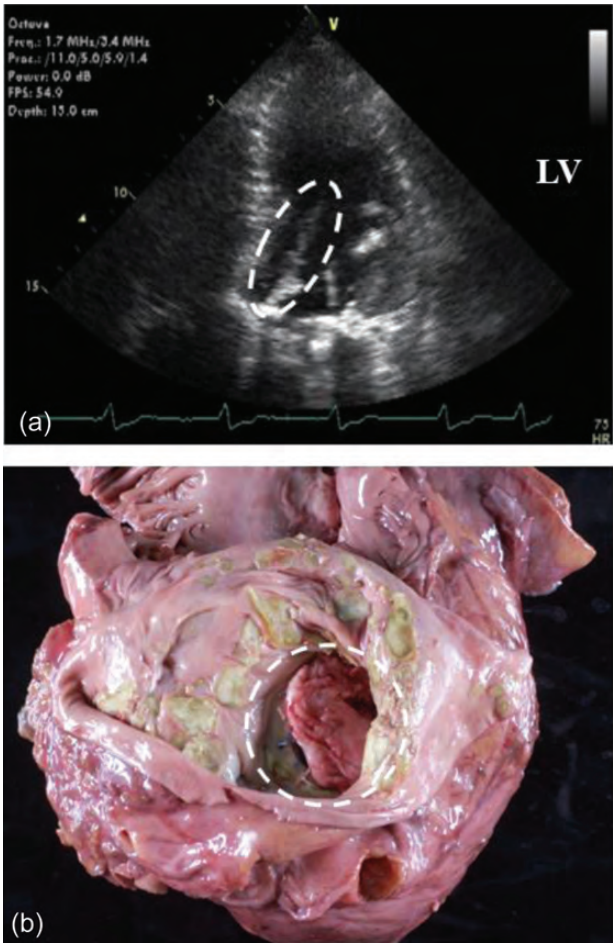

Fig. 2. Vegetation and thrombus in the patient's heart. Dotted circles represented the lesions in echocardiogram (**a**) and autopsy (**b**).

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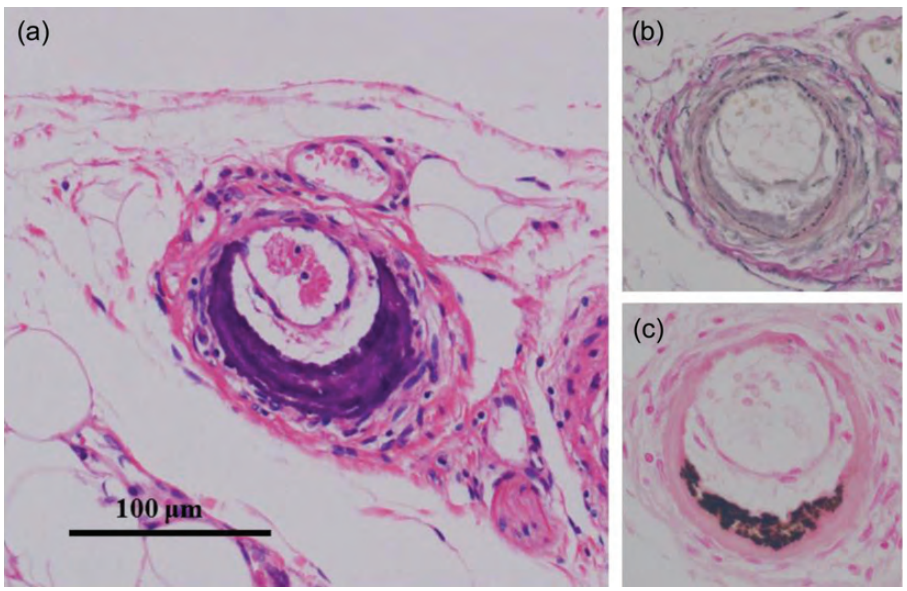

Fig. 3. Microscopic findings of vascular lesions of calciphylaxis. H–E staining showed media with concentric intimal hypertrophy. (**a**) EVG staining also showed the same findings. (**b**) Von Kossa staining showed the accumulation of calcium in the vessel wall and (**c**) another typical pathology of calciphylaxis.