Images in Nephrology
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Glomeruloid haemangiomas associated with generalized oedema in a patient with atypical POEMS syndrome

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A 55-year-old woman was admitted for evaluation of progressive generalized oedema and mild kidney dysfunction (serum creatinine 1.12 mg/dL; blood urea nitrogen 45 mg/dL). On admission, generalized oedema, transudative pleural effusion, collapsed inferior vena cava and low fractional excretion of sodium (0.14%) suggested severe volume depletion due to plasma extravasation; however, its aetiology could not be attributed to decreased colloid osmotic pressure (serum total protein 6.4 g/dL). Further examination of the patient revealed hepatosplenomegaly, slowing of nerve conduction velocities, albuminocytological dissociation in the cerebrospinal fluid, and multiple cherry-red papules on the neck and chest (Figure 1). These features were suggestive of polyneuropathy, organomegaly, endocrinopathy, monoclonal gammopathy and skin changes (POEMS) syndrome, also called osteosclerotic myeloma or Crow–Fukase syndrome. However, this case did not entirely fulfil the diagnostic criteria because monoclonal plasma cell disorders were not detected [1]. The skin biopsy revealed lesions consistent with glomeruloid haemangiomas (Figure 2), which is considered to be a specific marker for POEMS syndrome [2]. Furthermore, the serum level of vascular endothelial growth factor (VEGF), which is involved in the pathogenesis of POEMS syndrome [1], was extremely elevated to 16 300 pg/mL. These findings led us to make a diagnosis of atypical POEMS syndrome and speculate that microvascular hyperpermeability induced by excess VEGF resulted in generalized oedema and effusions associated with severe volume depletion [3,4]. In patients with oedema and effusions of unknown cause, one should consider the possibility of disorders associated with VEGF overproduction including POEMS syndrome.

References


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Fig. 1. Multiple eruptive cherry-red papules on the neck.

Fig. 2. (a) Dermal lesions showing vascular structures with capillary loops, resembling renal glomeruli (haematoxylin–eosin stain). (b) Endothelial cells stained for CD 31.

A specific marker for POEMS syndrome