



Glomeruloid Hemangioma as a Marker for the Early Diagnosis of POEMS Syndrome

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Dear Editor:

POEMS syndrome is an acronym for “polyneuropathy, organomegaly, endocrinopathy, M protein or monoclonal gammopathy, and skin changes,” and was first described by Bardwick in 1980¹. The cutaneous features of POEMS syndrome are common, including hyperpigmentation, hemangioma, hypertrichosis, white nails, sclerodermoid thickening, flushing, and clubbing^{1,2}. In particular, glomeruloid hemangioma is a well-documented histopathologically distinctive cutaneous vascular neoplasm associated with POEMS syndrome³. Here, we report a case of glomeruloid hemangioma as a marker for the early diagnosis of POEMS syndrome.

A 51-year-old man was consulted from the department of neurology because of a 1-year history of multiple eruptive papulonodular lesions on his trunk, with symmetrical progressively aggravating lower-limb weakness and numbness for suspected chronic inflammatory demyelinating polyradiculoneuropathy. A nerve conduction study revealed sensory-motor polyneuropathy in both lower limbs. He had no medical history except for hypothyroidism. On physical examination, multiple red papules and polypoid nodules, 2~6 mm in diameter, were found scattered on his trunk, and brownish hyperpigmentation with hypertrichosis were noted on his both forearms (Fig. 1A, B). He also showed clubbed fingers (Fig. 1C) and hardening of skin surfaces similar to scleroderma. Histopathologic examination demonstrated a dermal proliferation of capillary loops and vascular channels resembling renal glomeruli with eosinophilic globules within endothelial cells that were positive for periodic acid-Schiff stain (Fig. 2). The re-

sults of laboratory tests were all within the reference ranges and there were no evidences of systemic organ involvement. However, serum protein electrophoresis showed a light M-spike (2.1% of gamma) and immunofixation electrophoresis identified a monoclonal immunoglobulin A-lambda paraprotein, which is known to be the most common subtype in POEMS syndrome. In addition, plasma vascular endothelial growth factor (VEGF) level was 213 pg/ml (cut-off, 200 pg/ml). On the basis of these findings, POEMS syndrome was diagnosed and the patient was transferred to the department of hematology.

According to the annual updates in 2014, the mandatory major criteria for POEMS syndrome include polyneuropathy and monoclonal plasma cell proliferative disorder. For the diagnosis, one of the other major criteria (Castleman's disease, sclerotic bone lesion, and VEGF elevation) and one of the minor criteria (organomegaly, extravascular volume overload, endocrinopathy, skin changes, papilledema, and hematologic abnormalities) are additionally required to be present¹. However, making the diagnosis is a challenge particularly in the early phase of the disease because of the broad clinical spectrum based on multiorgan involvement. In the literature, glomeruloid hemangioma has been considered as a specific marker of POEMS syndrome since the study of Chan et al.⁴; however, some authors have reported a glomeruloid hemangioma that was not associated with POEMS syndrome⁵. There are controversies about the pathogenic relevance between glomeruloid hemangioma and POEMS syndrome; however when it appears, it mainly occurs in the early clinical course⁴. Therefore, dermatolo-

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Fig. 1. (A) Multiple, bright red, dome-shaped papulonodular lesions of 2~6-mm diameter were found scattered on the back and anterior chest. (B) Coarse hairs with surrounding brownish hyperpigmentation that were not previously present on the forearm, hand dorsum, and fingers were observed. (C) Schamroth's window test showed clubbed fingers with the loss of normal Lovibond angle.

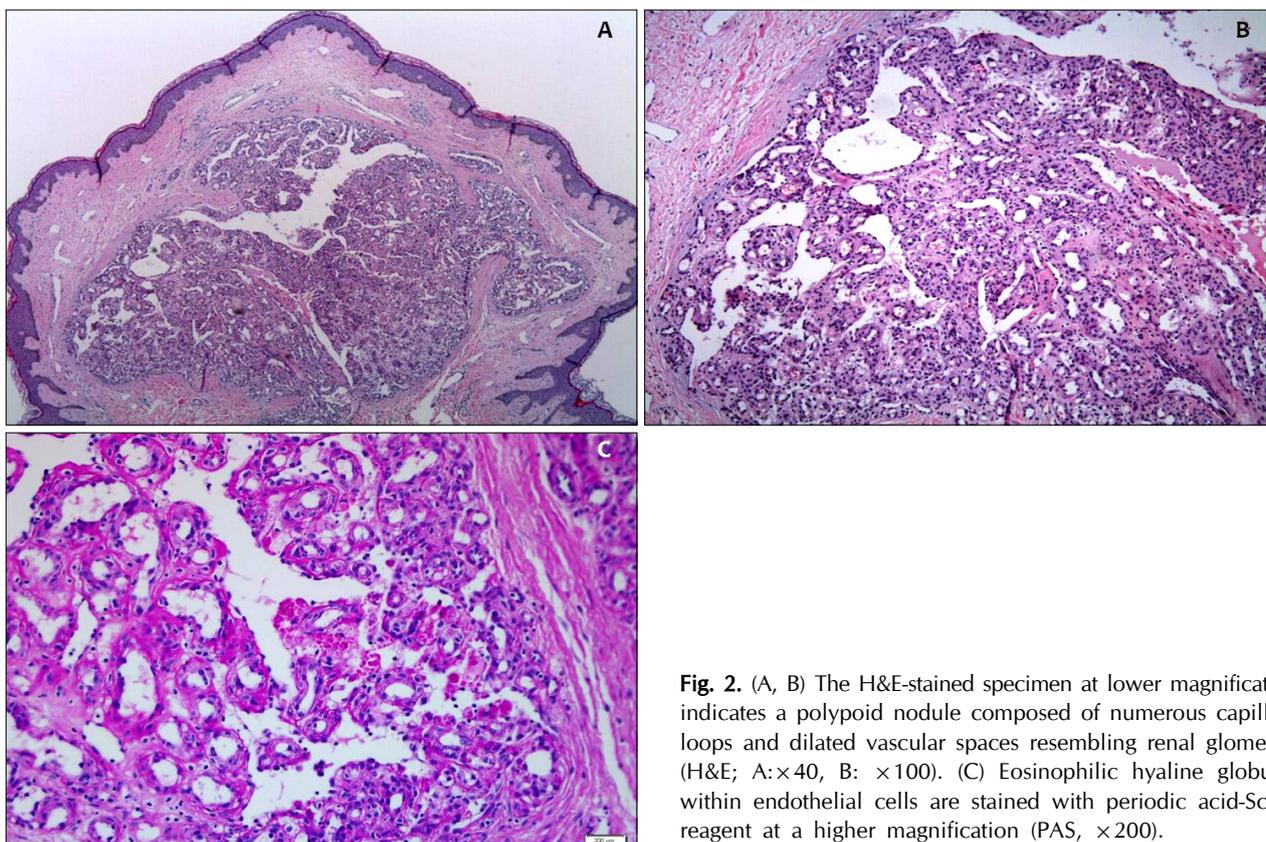


Fig. 2. (A, B) The H&E-stained specimen at lower magnification indicates a polypoid nodule composed of numerous capillary loops and dilated vascular spaces resembling renal glomeruli (H&E; A: $\times 40$, B: $\times 100$). (C) Eosinophilic hyaline globules within endothelial cells are stained with periodic acid-Schiff reagent at a higher magnification (PAS, $\times 200$).

gists should thoroughly evaluate eruptive hemangiomas when they are accompanied by polyneuropathy without other underlying causes. Early identification is important to allow a multidisciplinary management and avoid complications associated with thrombotic diatheses, including embolism, vascular dissection or necrosis, and volume overload including ascites, pleural effusion, and pericardial effusion¹.

CONFLICTS OF INTEREST

The authors have nothing to disclose.

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Hair Mineral Analysis in Children with Atopic Dermatitis

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Dear Editor:

Minerals and essential elements are important components of nutrition¹. These elements play crucial roles in the normal functioning of the immune system and antioxidant mechanisms which are related to the pathogenesis of atopic dermatitis (AD)². Previous studies have hypothesized that AD is associated with a non-specific decrease concerning trace metals^{3,4}. Furthermore, we previously reported that zinc (Zn) supplementation led to clinical improvement in AD patients with low hair Zn levels⁵. However, there is little data on other hair mineral levels in AD. Therefore, the aim of this study was to analyze the concentrations of trace elements in hair and to evaluate

their relevance to disease severity in children with AD.

A total of 66 children (37 boys, 29 girls; mean age, 5.88 years; range, 1~14 years) with confirmed diagnoses of mild to moderate AD (eczema area and severity index [EASI] scores < 26) were enrolled. A sex- and age-matched control group consisted of 25 children (15 boys, 10 girls; mean age, 6.12 years; range, 2~12 years) without dermatological disorders. The study protocol was approved by the ethics committee at Hanyang University Seoul Hospital (IRB no. 2011-R-34).

Participants were asked not to chemically process their hair for at least 8 weeks prior to mineral analysis. Mineral measurements were performed using a microwave tem-

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