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【 PICTURES IN CLINICAL MEDICINE 】

Glomeruloid Hemangioma in a Patient with TAFRO Syndrome

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Key words: glomerulus, immunoglobulin, POEMS syndrome, red papule, thrombocytopenia, vascular endothelial growth factor

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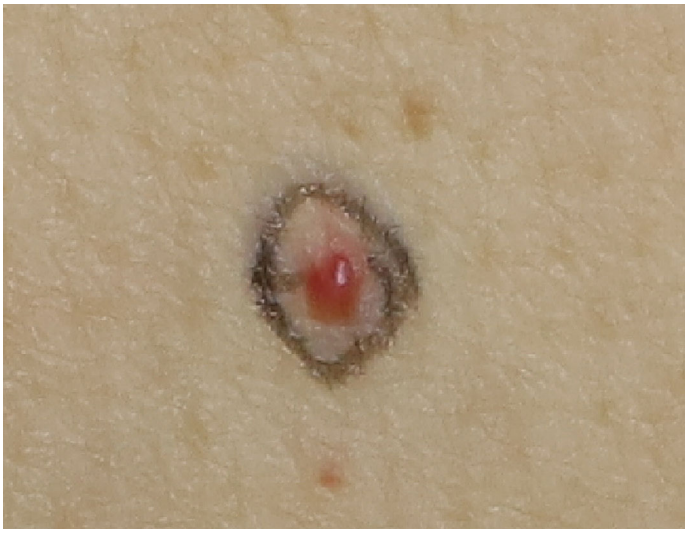

Picture 1.

A 61-year-old man developed fever, dyspnea, and painful abdominal bloating. Multiple red, dome-shaped papules were observed on his abdomen (Picture 1). He had thrombocytopenia (platelet count, 7×10^3/μL), and computed tomography revealed a large amount of pleural effusion and ascites, lymphadenopathy, and hepatomegaly. Although IgG-κ-type M protein was detected in the patient's serum, no endocrine abnormalities, polyneuropathy, or skin changes (e.g., hyperpigmentation) indicative of polyneuropathy, organomegaly, endocrinopathy, monoclonal protein, skin changes (POEMS) syndrome were apparent. Bone marrow biopsy revealed reticulin myelofibrosis without diffuse infiltration of plasma cells. Skin biopsy of a red papule revealed coiled aggregates of capillaries in the upper dermis with eosinophilic granules, resembling renal glomeruli (Picture 2a, b). These eosinophilic granules were positive for periodic acid-Schiff and kappa and lambda light chains (Picture 2c-e). The patient's serum vascular endothelial growth factor (VEGF) level was 131 pg/mL (normal, <38.3 pg/mL). The patient was diagnosed with thrombocytopenia, anasarca, myelofibrosis, renal dysfunction, and organomegaly (TAFRO) syndrome. The dyspnea and painful abdominal bloating were rapidly exacerbated and mechanical ventilation was required. Intravenous methylprednisolone pulse therapy with cyclophosphamide and plasma exchange allowed weaning from mechanical ventilation and improved the clinical manifestations. Glomeruloid hemangioma is considered a specific dermatopathological marker for POEMS syndrome, but has also been reported in patients with TAFRO syndrome (1, 2). The overproduction of VEGF and eosinophilic hyaline globules

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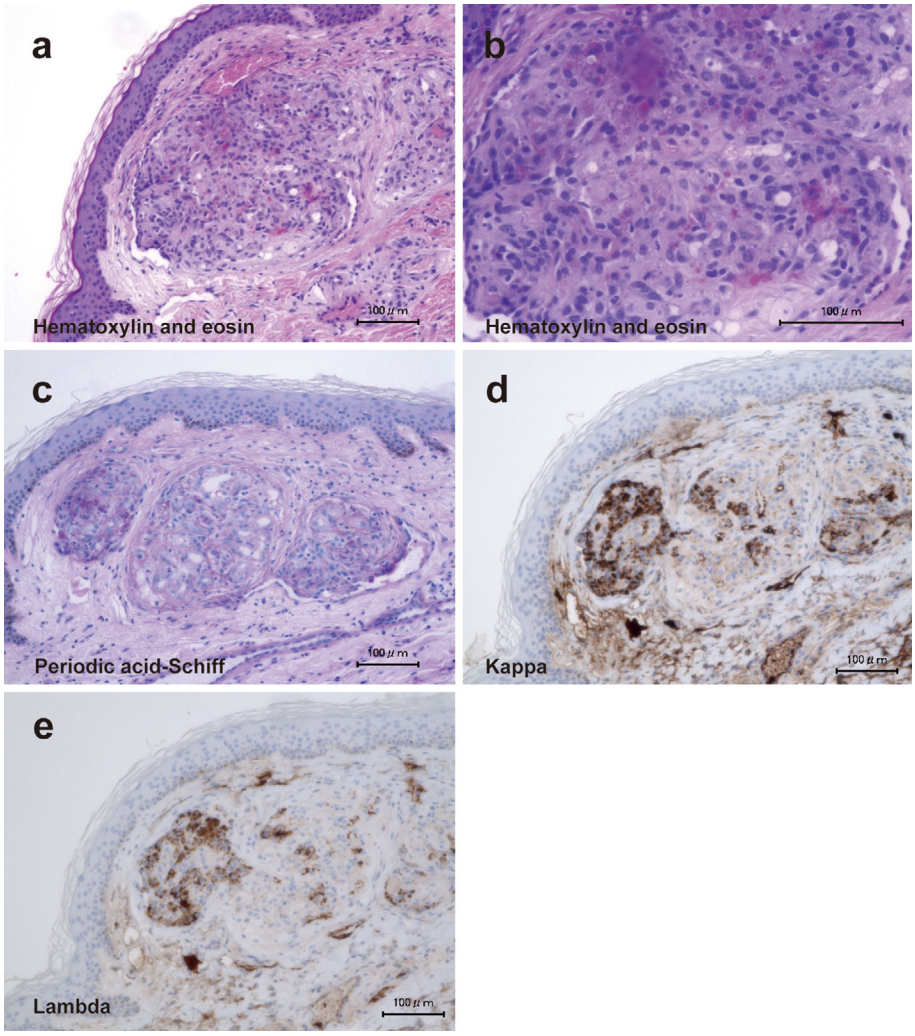

Picture 2.

containing immunoglobulins are speculated to be involved in the pathogenesis of glomeruloid hemangioma (1, 2). Although cutaneous manifestations of TAFRO syndrome have not been fully reported, glomeruloid hemangioma may be a diagnostic clue for TAFRO syndrome.

Written informed consent was obtained from the patient by the corresponding author. The signed consent forms have been retained by the corresponding author. We anonymized the patient's details as much as possible.

The authors state that they have no Conflict of Interest (COI).

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