中文題目: EB 病毒感染誘發的低補體性風疹塊樣血管炎症候群合併急性呼吸窘迫症候群

英文題目：Acute respiratory distress syndrome in a man with Epstein-Barr virus infection-induced hypocomplementemic urticarial vasculitis

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Introduction:

Hypocomplementemic urticarial vasculitis syndrome (HUVS) is a rare form of vasculitis with variable manifestations such as arthralgia, rashes, uveitis, and glomerulonephritis.[1] Pulmonary manifestations of the disease, such as pulmonary hemorrhage, could be fatal.[2] Because HUVS may manifest as various constitutional symptoms, it is difficult to diagnose the disease in the beginning. A carefully obtained history and thorough physical examination, followed by relevant laboratory investigations and histological examination are important for the diagnosis of the disease.

Case report:

A 45-year-old man, a non-smoker, presented with a 6-month history of recurrent non-pruritic macular skin rash and abdominal pain, and was admitted with 1 day's history of fever and myalgia. On examination, he was febrile (39.5°C) and had maculopapular rashes on the trunk and all extremities . A complete blood cell count yielded normal results for white blood cell count with eosinophilia (10.2%). Four days after admission, he developed progressive shortness of breath with desaturation. A computed tomography scan image of the chest revealed diffuse interstitial thickening with patchy ground-glass opacities. Endotracheal intubation was performed because of acute respiratory distress syndrome.

His serum complement levels were low, with a C3 level of 83.1 mg/dL and a C4 level of 9.9 mg/dL. Viral serologic investigations yielded negative results for hepatitis B and C, human immunodeficiency virus, influenza virus, and *Mycoplasma*. Serum polymerase chain reaction yielded positive quantitative result (28,829 copies/ml) for Epstein-Barr virus (EBV). Histologic examination of a skin biopsy specimen revealed mild perivascular leukocytoclastic infiltration in the upper dermis of the skin tissue . The immunohistochemical staining pattern was consistent with urticarial vasculitis. A diagnosis of HUVS was made and he recovered after a 1-week treatment course of intravenous methylprednisolone.

Discussion:

HUVS is an immune complex-mediated or type III hypersensitivity reaction.[1] Several pathogens such as hepatitis A, B and C viruses, EBV, and *Mycoplasma pneumoniae* have been implicated in the disease.[3,4] The diagnostic criteria for HUVS are classified into major criteria (chronic urticarial exanthema and hypocomplementemia) and minor criteria (leukocytoclastic vasculitis, arthralgia or arthritis, uveitis or episcleritis, glomerulonephritis, recurrent abdominal pain, and a positive C1q precipitin test).[5] A diagnosis is made based on fulfillment of major criteria and at least two minor criteria.

Pulmonary involvement in HUVS mainly presents as dyspnea, asthma, hemoptysis, cough, and chronic obstructive pulmonary disease.[2] Chronic obstructive pulmonary disease with basilar emphysematous radiologic changes occurs in half of patients with HUVS.[2] Although HUVS is associated with a low mortality rate, a previous study revealed a high mortality rate among patients presenting with respiratory failure.[2] Glucocorticoids and immunosuppressive therapy are recommended for the treatment of HUVS.[2, 3]

This patient presented with the typical manifestations of hypocomplementemic urticarial vasculitits accompanied with a fatal complication, acute respiratory distress syndrome. EBV was identified as the causative organism in this case. Although he developed acute respiratory distress syndrome, the symptoms were alleviated after treatment with methylprednisolone. The early diagnosis and initiation of treatment were keys to the successful treatment of this patient.

Reference:

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