An Unusual Case of Wegener's Granulomatosis

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Dear Sir,

Wegener's granulomatosis (WG) is a rare disease characterized by disseminated gran-ulomatous vasculitis which primarily affects upper and lower respiratory tract and the kidneys. Other organ systems can also be affected, including eyes, ears, skin, joints, heart, and the nervous system.

The disease runs a rapidly progressive course which usually leads to death within a few months, unless treated with immunosuppressive therapy [1]. We present a patient with a massive solitary pulmonary nodule who developed aggressive systemic features of WG and was treated by surgical resection.

A 49-year-old man was admitted to the hospital with weight loss, arthralgia, and weakness. His physical examination was normal. On chest X-ray, a round, well-arranged parenchymal mass, 12 cm in diameter, was found in the right middle lobe (fig. 1). Right middle lobectomy was performed with the diagnosis of a hydatid cyst made by the Department of Thoracic Surgery, but pathological examination of the material revealed a widespread inflammatory process with abscess formation, but without any evidence of a hydatid cyst. On the 13th day after the operation palpable purpura

characterized by bullae was detected, symmetrically on the feet and ankles of the patient.

These lesions were also present on hands, hip, sacrum, scrotum, and in mouth. Biopsy material obtained from the lesions on the feet revealed leukocytoclastic vasculitis. Oliguria and toxic retention developed. The patient was transferred to the Division of Nephrology. Biopsy material from the kidney revealed glomerulosclerosis and crescent formation. The laboratory data were as follows: urea 70 mg/dl, creatinine 5.6 mg/dl, K 6.4 mEq/l, Ca 7.8 mg/dl, P 8.1 mg/dl, hemoglobin 10.1 g/dl, white blood cells 28,000/

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Fig. 1. Round, well-arranged parenchymal mass in the right middle lobe.

Fig. 2. In the lung tissue, a necrotizing granuloma characterized by necrosis, surrounded by palisadic epithelioid histiocytes (on the left).

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mm3 (with a left shift), erythrocyte sedimentation rate 120 mm/h. The urinary sediment revealed 20 erythrocytes and 6-7 leukocytes (no eosinophils) and a few casts of erythrocytes.

Ultrasonographic examination of the abdomen revealed an increase in the cortical echogenicity and enlargement of both kidneys. All serum complement and immuno-globulin levels were normal and rheumatoid factor, antinuclear antibody, and anti-DNA were negative. The patient was positive for classic antineutrophil cytoplasmic autoanti-bodies. Therefore, in the light of the above findings, WG was diagnosed and treated with prednisolone and cyclophosphamide. Hemo-dialysis was instituted to treat acute renal failure. On the 33th day of his hospitaliza-tion, sepsis and disseminated intravascular coagulation developed, and the patient died. Postmortem examination of the lung revealed necrotizing granulomas and vasculitis with a widespread inflammatory process (fig. 2). These findings were consistent with the diagnosis of WG.

WG is a rare disease characterized by a generalized necrotizing vasculitis. The presence of circulating immune complexes and response to immunosuppressive agents make the autoimmunity a possible mechanism [2-4]. WG involves mostly the respiratory tract and the lungs. Lung infiltrates are typically multiple, nodular, bilateral lesions that frequently cavitate. The progressive form of this disease involves the lungs first and then generalizes to kidneys and skin and may result in a fatal outcome. As a sensitive test, classic antineutrophil cytoplasmic autoantibodies can be used for the diagnosis of WG [5]. Finally, we propose that in the differential diagnosis of a giant solitary nodule of the lung, as we have presented in this case, WG should be taken into consideration.

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