Histiocytosis X and Glomerulonephritis: A Case Report

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Dear Sir,

Recently, new types of glomerulopathies which are thought to develop secondary to different types of pathologies, have been reported. Since diagnosis of the primary cause is difficult, the definition of secondary glomerulopathy is crucial. Histiocytosis X is characterized by solitary or multifocal bone lesions. It is also called Langerhans cell histiocytosis or eosinophilic granuloma. Radiologically, a lytic destructive lesion is seen with no clear margins [1]. We present a case hospitalized with a prediagnosis of glomerulonephritis in whom afterwards eosinophilic granuloma in the costosternal region was detected. We would like to draw attention to this case from the point of coexistence of both disease and the outcome.

A 34-year-old married farmer was referred to the hospital, with 10 days’ history of fatigue, loss of appetite, high fever, headache, and vertigo. On admission to the hospital, he had signs of decreased urinary output. His blood pressure was 120/60 mm Hg, pulse rate 94/min, temperature 37 °C. Pathological findings on physical examination revealed edema on the eyelids and in the pretibial region. A rubbery, tender, immobile 4 × 4 cm mass lesion located on the right lower sterno-costochondral joint. The overlying skin was hyperemic and elevated by about 1 cm by the mass.

Laboratory findings on admission were as follows: Hb: 10.1 g/dl; Hct: 30.8%; blood leukocyte count: 16,300/mm3; BUN: 68 mg/dl; creatinine: 4.1 mg/dl; Na: 145 mEq/l; K: 5.2 mEq/dl; C3: 32 mg/dl; C4: 17 mg/dl. Anti-nuclear antibody, anti-DNA antibody and rheumatoid factors were negative. Urine gave (+++) test for protein, and urinary sediment contained 1-2 granular casts and 15-20 erythrocytes on microscopic examination. 24-hour protein loss in urine was 1.2 g. Ultra-soundographically, sizes of the right and left kidneys were 120 × 66 × 59 and 140 × 70 × 57 mm, respectively. Cortical thickness was 19 mm on the right and 20 mm on the left. Parenchymal echoes were at grade I.

On the 3rd day of hospitalization percutaneous renal biopsy was performed. In histopathological examination of the specimen crescents were seen in approximately 50% of the
glomeruli. Extensive interstitial infiltration of mononuclear cells was seen (fig. 1). On the 5th day, oligoanuria developed. BUN: 177 mg/dl; creatinine: 6.7 mg/dl; Na: 141 mEq/l; K: 6.8 mEq/l; Hct: 23.9%; leukocyte count: 13,300/mm³. Through a subclavian catheter, hemodialysis was performed and after 6 sessions the general condition of the patient improved. Two units of erythrocyte suspensions were given. Postdialysis serum BUN and creatinine levels were 57 and 2.4 mg/dl. The mass located at the lower sternal border was excised totally. Microscopic examination showed a typical granulomatous formation with histiocytosis, and infiltration of matured eosinophils and lymphocytes (fig. 2). Langerhans cell histiocytosis may present with different clinical manifestations. Unifocal bone lesions are mostly located at the cranium, mandible, ribs, femur, scapula and vertebrae [2]. Histiocytosis may lead to different types of endocrinopathies, owing to infiltration into the hypothalamus and the hypophysis. Diabetes insipidus, growth retardation, hypothyroidism, portal cirrhosis, intrahepatic cholestasis as well as involvement of lymph nodes, spleen, lungs and bone marrow may occur [3]. Coexistence of histiocytosis and nephropathy has not been reported so far. In our case, it is not obvious whether the two diseases are coexisting or not. Suppressor T-lymphocyte (CD8+) insufficiency due to abnormal thymic function resulting in an increase in T4/T8 is supposed to play a role in the etiopathogenesis of histiocytosis X [4]. It is known that the CD4+/CD8+ ratio is increased in some types of membranous and mesangial proliferative glomerulonephritis and in IgA nephropathy, but this finding is not clear for crescentic glomerulonephritis. Another interesting point is the rapid recovery of renal function after excision of the lesion. It is known that rapid-

Fig. 1. Sclerotic change of glomerulus with fibrocellular crescents and severe interstitial inflammation. Light microscopy. HE. × 200.

Fig. 2. The basic histologic lesion of Langerhans cell histiocytosis. Collections of histiocytes and variable numbers of mature eosinophils and lymphocytes. HE. × 400.

Rapid progressive glomerulonephritis is curable when the underlying cause is detected [5]. In our case, categorized as type II crescentic glomerulonephritis, the role of granuloma excision and the improvement of symptoms and signs is controversial. As a conclusion, we cannot express with certainty that in our case coexistence of histiocytosis and renal pathology is incidental, and it is not obvious whether excision of eosinophilic granuloma contributed to the healing of crescentic glomerulonephritis.

References

Nephron 1996;73:329-330
Duranay/Akbas/Erbilen/Bali/Sahin/Arab/ Kuzu