A rare case of dermatomyositis associated with immune-complex type glomerulonephritis, idiopathic thrombocytopenic purpura, pulmonary fibrosis and lung cancer

Sir,
Renal involvement is a poor prognostic factor in systemic autoimmune diseases. However, such involvement is uncommon in polymyositis/dermatomyositis (PM/DM). We report a rare case of dermatomyositis associated with immune-complex type glomerulonephritis, idiopathic thrombocytopenic purpura, pulmonary fibrosis and lung cancer.

A 49-year-old Japanese male was referred to the University Clinic and Hospital for further evaluation of sudden onset of high fever, arthralgia and myalgia of extremities, erythema and swelling of eyelids. The past history, family history and lifestyle were not remarkable. Physical examination on admission revealed apparent heliotrope erythema on edematous eyelids, marked Gottron sign on finger joints, fine crackles audible on chest auscultation, and muscle weakness in the upper and lower extremities.

Initial investigation demonstrated positive tests for inflammation; leukocytes 6,100/µl (band 13%, segmented 73%, and lymphocytes 6%), C-reactive protein 1.9 mg/dl (normal < 0.2), erythrocyte sedimentation rate 30 mm/hr, high serum levels of AST 564 IU/l, LDH 6,100 IU/l (band 13%, segmented 73%, and lymphocytes 6%), C-reactive protein 1.9 mg/dl (normal < 0.2), erythrocyte sedimentation rate 30 mm/hr, high serum levels of AST 564 IU/l, LDH 6,100 IU/l (normal < 0.2), erythrocyte sedimentation rate 30 mm/hr, high serum levels of AST 564 IU/l, LDH 6,100 IU/l (normal < 0.2), erythrocyte sedimentation rate 30 mm/hr, high serum levels of AST 564 IU/l, LDH 6,100 IU/l (normal < 0.2), erythrocyte sedimentation rate 30 mm/hr, high serum levels of AST 564 IU/l, LDH 6,100 IU/l (normal < 0.2), erythrocyte sedimentation rate 30 mm/hr, high serum levels of AST 564 IU/l, LDH 6,100 IU/l (normal < 0.2), erythrocyte sedimentation rate 30 mm/hr, high serum levels of AST 564 IU/l, LDH 6,100 IU/l (normal < 0.2), erythrocyte sedimentation rate 30 mm/hr, high serum levels of AST 564 IU/l, LDH 6,100 IU/l (normal < 0.2), erythrocyte sedimentation rate 30 mm/hr, high serum levels of AST 564 IU/l, LDH 6,100 IU/l (normal < 0.2), erythrocyte sedimentation rate 30 mm/hr, high serum levels of AST 564 IU/l, LDH 6,100 IU/l (normal < 0.2), erythrocyte sedimentation rate 30 mm/hr.

Chest radiography and computed tomography demonstrated acute pneumonia and fibrosis. High dose methylprednisolone was administered intravenously for 3 consecutive days, followed by oral prednisolone 80 mg/day. However, the severity of dyspnea worsened and the patient died of respiratory distress two weeks after admission.

An autopsy was performed. Degenerative and fibrotic changes in the skeletal muscles as signs of chronic inflammation, and epidermal thinning confirmed the initial diagnosis of dermatomyositis. Histopathologic examination of the kidney under light microscopy showed minor glomerular abnormalities with adhesion of glomerulus to the capsule (Fig. 1a). However, fluorescent immunostaining and electron microscopic examinations revealed heavy deposits of immunoglobulins and complement deposits at subendothelial regions (Fig. 1b and 1c). Pathology of the lungs demonstrated fibrosis and bronchopneumonia, and unveiled an occult squamous cell carcinoma in the bilateral upper lobes, but not in other organs or tissues.

Renal cell carcinoma concurrent with PM/DM as an autoimmune disease-associated malignancy, and acute renal failure due to massive rhabdomyolysis of PM/DM, have been occasionally reported. However, glomerular involvement is rare in PM/DM. Dyke et al. (1) reported four cases of polymyositis with proteinuria, and emphasized focal mesangial proliferation as a characteristic feature of the renal involvement. However, not only mesangial proliferation but also wide ranges of pathological findings from minor glomerular abnormality to membranous nephropathy and crescentic glomerulonephritis have been observed (1-10). In those reports, fluorescent immunostaining was performed in 12 patients, and immunoglobulins and/or complements were demonstrated in 9 out of the 12 samples. Together with the wide spectrum of histopathologic findings, these suggest that immune complexes play pivotal role in the pathogenesis of the majority of PM/DM-related nephropathy.

In summary, glomerulonephritis is an infrequent complication of PM/DM. It could manifest in a wide spectrum of histopathologic changes, and be caused by immune complex associated autoimmune mechanism, similar to lupus nephritis.

Fig. 1. Histopathological examination of the kidney from autopsy specimens. (a) Minor glomerular abnormalities with adhesion of glomerulus to the capsule (PAS staining, orig.mag. x100). (b) Immunostaining of the mesangial region and glomerular basement membrane by immunoglobulins G, A, and M, and complements 1q and 3 (immunofluorescence for IgA, orig.mag. x40). (c) Electron micrograph of glomerulus. Electron dense deposits (arrows) can be seen in the mesangial and subendothelial regions (x2,900).

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Letters to the Editor

References


Erratum

In the Letter to the Editor "Familial Mediterranean fever: Is low mortality from tuberculosis a specific advantage for MEFV mutation carriers? Mortality from tuberculosis among Muslim, Jewish, French, Italian and Maltese patients in Tunisia (Tunisia) in the first half of the 20th century" by D. Cattan published in the supplement issue Behcet's Disease and Familial Mediterranean Fever no. 3 (Clin Exp Rheumatol 2003, vol. 21, no. 4, suppl. no. 30, pgs. S53-S54) there were two errors in the data reported from the "Statistiques Sanitaires et Démographiques; Régence de Tunis; Protectorat Français". In Tunis the number of deaths from tuberculosis per 100 deaths in Muslims was 13.6 (7.3 – 18.2) rather than 13.6 (7.3 – 10.2). The contribution of tuberculosis to deaths in Tunis was 79.3% (57 – 79.3) rather than 77.3% (57 – 79.3).

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