# Dermatomyositis with membranous nephropathy

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SUMMARY: Soylu A, Kavukçu S, Türkmen M, Sarıoğlu S. Dermatomyositis with membranous nephropathy. Turk J Pediatr 2001; 43: 143-145.

Dermatomyositis is the connective tissue disease with the least renal involvement. Although some renal findings like proteinuria, hematuria, pyuria, progressive renal insufficiency, and glomerular and tubular calcium deposits with arteriolar fibrosis have been described, glomerulonephritides have rarely been associated with dermatomyositis, especially in childhood cases. We describe a 10-year old boy with the clinical picture of dermatomyositis who underwent renal biopsy due to microscopic hematuria demonstrating membranous glomerulonephritis with C1q deposition. Children with "full-house" membranous glomerulonephritis with deposition of C1q and the other immunoglobulins have been reported to present in the future with the clinical findings of systemic lupus erythematosus. However, laboratory evaluation of our patient for systemic lupus erythematosus was negative at the present time. Thus, we think this case should be followed up closely with special attention to the possible clinical and laboratory findings of systemic lupus erythematosus.

*Key words: dermatomyositis, membranous nephropathy, systemic lupus erythematosus, childhood.*

Dermatomyositis (DM) is the connective tissue disease with the least renal involvement. Although DM is considered a necrotizing vasculitis, there is very little evidence to suggest that endothelial injury occurs in the kidney. However, some renal findings like proteinuria, hematuria, pyuria, progressive renal insufficiency, and glomerular and tubular calcium deposits with arteriolar fibrosis have been described[1]. Glomerulonephritis [membranous glomerulonephritis (MGN) specifically] has been associated with DM in only a few cases[1-3]. Full-house MGN has been suggested as a preceding marker of systemic lupus erythematosus (SLE) in childhood[4].

We describe a 10-year old boy with DM who developed microhematuria two months after the diagnosis of DM. Persistence of hematuria led to a percutaneous renal biopsy demonstrating C1q-positive MGN.

## Case Report

A 10-year-old boy presented with a two-month history of weakness and pain in his arms and legs and difficulty in walking. There were in addition erythema of his eyelids, nasal speech and difficulty in eating solid food. Personal and family histories were unremarkable. Physical examination revealed heliotropic rash (Fig. 1), weakness and tenderness in the proximal muscles of the extremities, and decreased deep tendon reflexes in the lower extremities. Laboratory analysis demonstrated Hb 11.9 g/dl, Hct 36%, WBC 16,900/mm$^3$, platelets 251,000/mm$^3$, MCV 81 fl; ESR 13 mm/h, ASO 85 IU/ml, CRP 5 mg/dl, RF (–); BUN 17 mg/dl, creatinine 0.4 mg/dl, SGOT 108 IU/ml, SGPT 26 IU/ml, and CPK 955 IU/ml. Pulmonary function tests showed moderate restrictive disease. Urinalysis revealed specific gravity 1030, pH 5.0, protein (–), blood (–), and 1-2 WBC/hpf. Electromyogram demonstrated diffuse muscle fiber involvement in the proximal muscles with intense spontaneous fibrillation indicating acquired inflammation. Muscle biopsy showed mononuclear cells infiltrating vessel walls within muscle and fat tissue and atrophic changes in muscle cells in some areas.

Treatment was started with prednisolone, 2 mg/kg/day in four divided doses, and the dose was decreased gradually after two months along with institution of physiotherapy, upon improvement of the active symptoms and signs of the disease. However, microscopic hematuria without proteinuria was noted in routine urinalysis

three months after the diagnosis. His blood pressure was normal. Urine culture was sterile and urinary system ultrasonography was normal. Complete blood count, erythrocyte sedimentation rate, C-reactive protein, C3, C4, and urinary calcium excretion were normal. ANA was negative. Persistence of microscopic hematuria led us to perform a renal biopsy six months after the diagnosis of DM. Histopathologic evaluation of the renal tissue revealed basal membrane thickening without mesangial proliferation, minimal interstitial fibrosis and tubular atrophy in light microscopy (Fig. 2). Direct immunofluorescent microscopy demonstrated 3+ membranous, fine granular, linear IgG, IgA and IgM deposition with minimal C3, C1q, fibrinogen and albumin deposition (Fig. 3). Thus, the diagnosis of MGN was made histopathologically.

Fig. 1. Heliotropic rash on the face of the patient.

Fig. 2. Slight basal membrane thickening in a glomerulus (periodic acid-Schiff PAS, x 200).

## Discussion

Glomerulonephritis has been described in only a few cases of DM[1-3,5]. Review of the literature revealed only one pediatric patient with MGN developing DM during follow-up. That child had developed proteinuria with nephrotic syndrome before the diagnosis of MGN. Our case is a 10-year old boy diagnosed as DM before he developed microscopic hematuria.

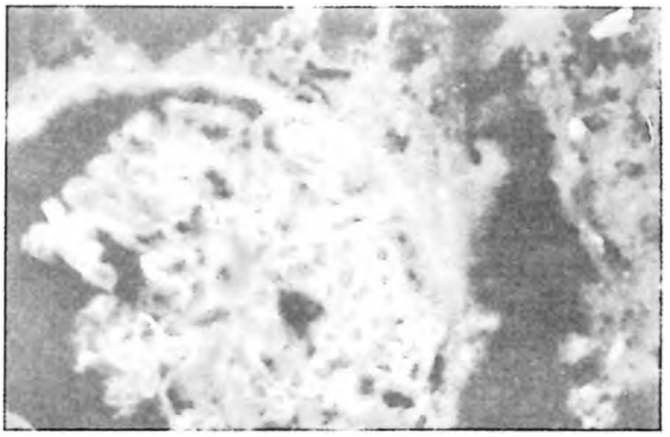

Fig. 3. Fine granular 3+ IgG depositon in the basal membrane (DIF, IgG, x 400).

Since renal involvement is so rare in DM, we reevaluated the patient to exclude other collagen vascular diseases such as SLE, but no clinical or laboratory evidence of SLE was detected. In addition, hypercalciuria could have been suspected as a cause of hematuria, since the patient developed hematuria during treatment with corticosteroid therapy. However, hematuria was detected during the dose reduction period of steroid therapy, and renal calcium excretion was determined to be normal in serial measurements. Seraches for the other causes of hematuria were also negative.

Persistence of hematuria without any known cause led us to perform a renal biopsy that demonstrated MGN. Lack of marked mesangial proliferation resembled idiopathic MGN histopathologically, not the membranous nephropathy of SLE. However, accumulation of C1q, a marker of renal tissue involvement in SLE, was noted. Presence of C1q-positive MGN in renal histopathology with the clinical findings of DM in this child suggested that our patient represented a case of seronegative SLE with renal involvement. It has been reported that children with "full-house" MGN with deposition of C1q and the other immunoglobulins could present in the future with the clinical findings of SLE, especially when cytoplasmic tubuloreticular inclusions are detected in electron microscopy[4,6]. Unfortunately, we could not evaluate the renal biopsy specimen by electron microscopy.

However, in view of the data mentioned above[4,6], although the clinical picture of our case is compatible with DM at the present time, other clinical and biological symptoms of SLE may appear in this patient in the future.

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