Is bullous skin lesion a risk factor for renal amyloidosis in patients with familial mediterranean fever?

Sir,

Familial Mediterranean fever (FMF) is an autosomal recessive disorder characterized by inflammation of the serous membranes. Clinical signs include fever, abdominal pain, arthritis, erysipelas-like erythema, and chest pain. The most important and feared complication of the disease is amyloidosis. Colchicine is useful to prevent attacks and amyloidosis.[1]

A 60-year-old male patient presented with chest pain, swelling of the feet, and recurrent fever or 15 days and his medical history was unremarkable except for hyperlipidemia, asthma treated with the theophylline 400 mg/day, atorvastatin 20 mg/day, and montelukast 4 mg/day. On physical examination, he had +2 pretibial edema bilaterally and 4-5 bullous skin lesions measuring 1 cm × 1.5 cm on his back. Investigations showed hemoglobin 12.2 g/dl, leukocyte count 10.620/mm^3, alanine transaminase, 7 U/L, gamma glutamyl transferase 14 U/L, total bilirubin 0.7 mg/dl, albumin 2.3 g/dl, globulin 2.4 g/dl, creatinine 1.24 mg/dl, urea 30 mg/dl, C-reactive protein 44.76 mg/L, and 2840 mg/day proteinuria. Kidney biopsy was suggestive of amyloid, and immunohistochemical staining was positive for amyloid A, suggesting secondary amyloidosis. Genetic analysis revealed the presence of homozygous R202Q mutation in MEFV gene. He was started on colchicine, which led to regression of skin lesions. However, the serum creatinine level progressed to 5.39 mg/dl within 3 months, and he was started on hemodialysis. The patient is still follow-up in our clinic.

The MEFV gene is responsible for FMF. The mutations of the gene may be responsible for fever, skin lesions, and other additional clinical findings. The presence of homozygous M694V mutations, sex, intermarriage, arthritis, and resistant microalbuminuria are risk factors for amyloidosis.[2] The most characteristic cutaneous manifestation of FMF is erysipelas-like erythema. Other skin manifestations are urticaria, non-specific purpura, psoriasis, and erythema nodosum.[3] Bullous skin lesions may appear as a rare skin lesion.[4] Histopathological findings of the peritoneal cavity and synovial biopsy specimens are similar with the dermis during attacks. The association between other skin lesions except for erysipelas-like erythema and presence of homozygous M694V mutations is still controversial.[2] In the literature, a patient with FMF who was stable since more than 14 years reported that the patient progressed to renal failure with the skin lesions.[5] The incidence of renal amyloidosis is 20-25% in patients with untreated FMF. Although the presence of gene mutations are common in amyloidosis and skin findings, bullous lesions may accelerate amyloid deposition.

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