

(близких родственников), хорошо функционируют у 90–95% пациентов в течение 1 года и у 85–90% в течение 2 лет. Известны сроки выживания больных с пересаженными почками более 20 лет.

КЛЮЧЕВЫЕ слова: трансплантация почки, врожденный ЦМВ-ассоциированный нефротический синдром, детский возраст.

CASE REPORT: TWO FAMILIAL MEDITERRANEAN FEVER PHENOTYPE 2 CASES PRESENTED WITH RENAL FAILURE CAUSED BY AMYLOIDOSIS

Ebru UZ¹, Cemaleddin ÖZTÜRK², F.Betül ASAN², Şükran ERTEN³

¹ *Yildirim Beyazıt University Faculty of Medicine Department of Nephrology*

² *Yildirim Beyazıt University Faculty of Medicine Department of Internal Medicine*

³ *Yildirim Beyazıt University Faculty of Medicine Department of Romatology*

Familial Mediterranean Fever (FMF) is a genetic, inflammatory disease. Familial Mediterranean Fever is presented with fever attacks, serositis, arthralgia, arthritis and local inflammatory symptoms and findings. Two FMF cases are presented with amyloidosis and renal failure.

CASE 1: A 36 years old man admitted to the hospital with nausea and vomiting. He had a history of deep venous thrombosis and left nephrectomy after a traffic accident six years ago. Physical examination revealed cachectic appearance, three positive pitting edema in pretibial and sacral areas and the remainder examination was normal. Biochemical analysis revealed serum BUN 131,7 mg/dl and serum creatinin 8,75 mg/dl. Arterial blood gases examination revealed Ph: 7,1 and HCO₃ 3 mEq/L. The patient has been started on dialysis. The patient had 4 gr/day proteinuria with serum albumin level 1,4 gr/dl. Immune serologic laboratory markers were negative. Because of left nephrectomy history renal biopsy was contraindicated. Pathologic examination of rectum biopsy reveals type AA amyloidosis. We performed a molecular analysis for mutations in MEFV gene. The

results showed heterozygous E148Q mutation.

CASE 2: A 39 years old woman admitted to the nephrology department with nausea, vomiting and generalized edema. She had no history of disease. Biochemical analysis revealed serum albumin 2,2 gr/dl, serum creatinine 3,6 mg/dl and urine albumin 9 gr/day. Pathologic examination of renal biopsy revealed type AA amyloidosis. In molecular analysis there was no FMF gene mutation. The patient has been considered as type 2 FMF and started on routine hemodialysis programme

There was no etiologic cause for secondary amyloidosis in both cases

DISCUSSION: Familial Mediterranean Fever can be presented as different phenotypes.

Phenotype 1 is characterized with recurrent fever, peritonitis, synovitis, pleural effusion, pericarditis, orchitis, meningitis and amyloidosis.

Phenotype 2 is characterized with AA amyloidosis without symptoms.

Phenotype 3 is silent carrier of heterozygotes or homozygous MEFV gene mutations.

Physicians should keep in mind Phenotype 2 FMF in case there is no proteinuria and typical symptoms of the FMF.

Нефротический синдром в пожилом возрасте. Трудности постановки диагноза. Клинический разбор.

Молдахметова С. С., Туребеков Д. К., Ногайбаева А. Т.

ТОО «Медицинская клиника «Центр гемодиализа»», г.Астана, Казахстан

ГКП на ПХВ «Городская больница №1», г.Астана, Казахстан

АО «Национальный научный кардиохирургический центр», г.Астана, Казахстан

ВВЕДЕНИЕ. Первичные гломерулярные заболевания почек в пожилом возрасте встречаются редко, а вторичные гломерулонефриты требуют дифференциальной диагностики между паранеопластическим процессом и метаболическими