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Hemolytic Uremic Syndrome as a Cause of Adult Acute Renal Failure

Erişkin Akut Böbrek Yetmezliğinin Bir Sebebi Olarak Hemolitik Üremik Sendrom

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Abstract

Thrombotic microangiopathy is a microvascular occlusive disorder characterized by microangiopathic hemolytic anemia, thrombocytopenia, and variable signs of organ injury due to platelet thrombosis in the microcirculation. Regarding to the severe of brain or renal lesions, two clinical entities (pathologically similar but clinically different) are described: thrombotic thrombocytopenic purpura and hemolytic uremic syndrome. Thrombotic thrombocytopenic purpura usually affect adults, and is characterized by severe neurologic involvement in most cases, and variable renal involvement. Hemolytic uremic syndrome occurs in young children, and is characterized by acute renal failure, and absent or minimal neurologic abnormalities. In this case report, the clinical features, diagnosis, pathophysiology, and treatment of an adult with acute renal failure due to the thrombotic microangiopathy with hemolytic uremic syndrome are discussed.

Keywords: Acute Renal Failure, Thrombotic Thrombocytopenia; Hemolytic-Uremic Syndrome, İntensive Care.

Öz

Trombotik mikroanjiopati, mikroanjiopatik hemolitik anemi, trombositopeni ve çeşitli organ tutulumları ile karekterize mikrovasküler bir hastalıktır. Trombotik mikroanjiopati'de beyin veya böbrek tutulumunun ciddiyetine göre; patolojik olarak benzer, ancak klinik olarak farklı iki klinik tablo tarif edilmiş olup; bunlar trombotik trombositopenik purpura ve hemolitik üremik sendrom olarak adlandırılmıştır. Trombotik trombositopenik purpura daha çok erişkinlerde görülmektedir, nörolojik bulgular ön plandadır ve böbrek tutulumu hafiftir. Buna karşın hemolitik üremik sendrom, daha çok çocukluk çağında görülür, akut böbrek yetmezliği belirleyici özellik olup, nörolojik bulgular hafif veya yoktur. Bu olgu sunumunda erişkin akut böbrek yetmezliği gelişen trombotik mikroanjiopatiye bağlı hemolitik üremik sendromun klinik bulguları, tanısı, patofizyolojisi ve tedavisi tartışılmıştır.

Anahtar Kelimeler: Akut Böbrek Yetmezliği, Trombotik Trombositopeni, Hemolitik Üremik Sendrom, Yoğun Bakım.

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INTRODUCTION

The common findings of thrombotic thrombocytopenic purpura (TTP) and hemolytic uremic syndrome (HUS) are microangiopathic hemolytic anemia, intravascular thrombus/fibrin formation, organ dysfunction, and thrombocytopenia (1-3). TTP is one of the most common causes of acute renal failure in infants and children (4). It is rare in adults. HUS may also rarely occur following glomerular pathology. The underlying pathology is thrombotic microangiopathy accompanied by microvascular occlusive disorder of often capillaries or less frequently of arteries (3). Thrombotic thrombocytopenia purpora, which shares similar clinical features and pathologies, often causes cerebral ischemic lesions due to platelet aggregation but platelet-fibrin thrombus usually affect the kidneys in HUS (5, 6). However, TTP-HUS continue to be a life-threatening disease despite of the early treatment strategies. In this case report, the clinical features, diagnosis, pathophysiology, and treatment of an adult with acute renal failure due to the thrombotic microangiopathy (MA) with hemolytic uremic syndrome are discussed.

CASE REPORT

Table 1 presents laboratory findings of a 22-year-old female patient at emergency department admission and after treatment who was diagnosed with HUS and presented with abdominal pain, nausea, vomiting, diarrhea, confusion, anuria, hypotension and shock in our emergency department. She had been hospitalized 3 days ago due to diarrhea and had been discharged with antibiotic therapy. On the initial examination the patient was unconscious, intubated, hypotensive, and tachycardiac in the emergency department. The patient was transported to intensive care for taking care and treatment. We connected the patient to mechanical ventilator in spontaneous assisted breathing mode and performed full monitoring (ECG, temperature and oxygen saturation tests, and arterial monitoring). We also obtained all the cultures and pre-diagnosed the patient with sepsis. Within the first hour, we administered a broad-spectrum antibiotic therapy. The thorax tomography revealed haemathorax in right lung and the right chest tube was placed. As the patient had a hypotensive course, we started to administer dopamine infusion. The patient was then taken to the hemodialysis after hemodialysis catheter was placed. After the first ten days, the patient still could not be detached from the mechanical ventilation upon which we applied percutaneous tracheostomy with Griggs method. The parenteral nutrition that had started on hospitalisation was further supported with enteral nutrition on the second day. Then the parenteral nutrition was replaced by a 31-day enteral nutrition. The patient received eight sessions of hemodialysis and had to discontinue the session due to acute renal failure after plasmapheresis three times. The patient stayed in intensive care for 49 days and was transferred to the infectious disease department.

Table 1. The laboratory findings at emergency department admission and after the treatment.

	Before	After
Leucocytes (/mm^3)	13700	4300
Hemoglobin (gr/dL)	10,6	7,1
Haematocrit (%)	30	20
Platelet (/mm^3)	24000	106
CRP (gr/dL)	6,18	10,4
Glucose	100	95
LDH (U/L)	1760	670
Lipase U/L	16	17
Amylase U/L,	34	33
ALP U/L	46	
Urea (mg/dl)	40	59
Creatinin (mg/dl)	5,76	4,93
AST (U/L)	131	15
ALT (U/L)	131	12
GGT(U/L)	8	6
Albumin	2,8	2,5
Sodium (mEq/lt)	133	134
Potassium (mEq/lt)	2,8	3,6
Phosphor (mg/dL)	7,2	5
Total biluribin (mg/dL)	0,8	0,9
Direct biluribin (mg/dL)	0,4	0,5
PTT	13,6	10
aPTT sec (24-36)	34,4	25
Ptz	65%	60%
Fibrinogen mg/dL	302	200
D-Dimer mikrogFEU/mL	4,4	1
INR	1,09	0,9
Anti HBsAg IU/mL (micro)	19,5	
AntiHAVIgG	Positive	
Anti-HCV	Negative	
pH	7,36	7,36
pCO2 mmHg	32	36
pO2 mmHg	230	100
Saturasyon %	99	98
BE	-6,5	2,3

DISCUSSION

HUS is characterised by thrombocytopenia, acute deterioration in renal function, microangiopathic anaemia, and endothelial dysfunction. HUS is rare and serious disease with a variety of etiologies among adults. HUS is also one of the rare causes of acute renal failure in adults (7, 8). The chronic renal failure rate after HUS is between 40-60% (8-10). Our patient was diagnosed with HUS due to the presence of acute renal failure, thrombocytopenia, and microangiopathic haemolytic anaemia triad. Because of the presence of fever, diarrhea, hypotension, leukocytosis, and inotropic need, we diagnosed the patient with HUS-related sepsis. There was no underlying chronic diseases. Studies in human kidney have shown that cytotoxin often binds with renal tubular cells close to the glomeruli and that the cytokines produced by such cells may play a role in the pathogenesis of HUS (2).

Central nervous system and neurological dysfunction are common pictures in HUS (11-13). The severity of acute illness has been found to be associated with the first need of dialysis and symptoms of central nervous system (7, 11). Our patient had clouded consciousness and

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convulsions. Microthrombus in HUS patients constitute the most damage in the kidneys (2). In our case, too, there was acute renal failure. Our patient received 8 sessions of hemodialysis and underwent plasmapheresis three times. We administered fresh frozen plasma and regular antibiotic therapy. The patient eventually recovered from renal insufficiency. The literature also reports the case of a patient who developed HUS-related renal failure in the postpartum period and recovered with TDP, plasmapheresis, and hemodialysis (6,7).

In a cohort study, plasmapheresis administration has provided high response and survival rates (6). However, the literature also reports the case of a patient who, after undergoing plasmapheresis for 28 times, could not recover from renal failure and had to need renal raplacement therapy (14). Another study reports the case of a HUS patient accompanied by a secondary disease with increased mortality and recurrence (15,16).

As a result, HUS is a rare yet major cause of kidney failure in adults. Still, a convenient and fast fluid electrolyte therapy, antibiotics, and dialysis and plasmapheresis for patients with renal failure may reduce mortality.

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