DESCRIPTIVE ANALYSIS OF AN HISTORICAL COHORT OF PATIENTS WITH THROMBOTIC MICROANGIOPATHY (TMA)

T. Linares, A. García-Prieto, M. Goicoechea, F. Anaya, U. Verdalles, A. Santos, MS. García de Vinuesa, C. de Lucas, P. Rodriguez-Benitez, A. Luque y J.Luño. Department of Nephrology. Hospital General Universitario Gregorio Marañón, Madrid, Spain.

INTRODUCTION

Despite their diversity, TMA syndromes are united by common, defining clinical and pathological features that include microangiopathic hemolytic anemia, thrombocytopenia and organ injury. The classic TMAs are hemolytic uremic syndrome (HUS), thrombotic thrombocytopenic purpura (TTP) and secondary TMA.

METHODS

A retrospective study of a historic cohort of patients diagnosed with thrombotic microangiopathy (TMA) from 1984-2015 at a single center was performed.

RESULTS

96 patients were included: 63 adults (median age: 43.1±16) and 33 children (median age: 3.7±3.8). In adults the diagnosis was TTP in 23 cases, HUS in 14 cases and secondary TMA in 26 cases. In children 33 cases (100%) had been diagnosed as HUS.

In patients with HUS, the average age at the time of diagnosis was 35.5±11 years in adults and 3.7±3.8 in children. The most prevalent clinical symptoms were oliguria (p=0.002) and diarrhea (p=0.03) in HUS and petechiae in TTP (p=0.011). The degree of thrombocytopenia was most severe in patients with TTP. 89% of patients with HUS had thrombocypenia, being more severe in adults. ADAMTS13 testing was performed only in 12 patients. There were no statistically significant difference between adults and children in the hemoglobin level or in their renal function at the time of diagnosis. 70% of children and 50% of adults patients presented with diarrhea, but only 15 children and 2 adults had Shiga toxin test performed (5 children and 2 adults tested positive).

71% of adults and 9% of children were treated with plasma exchange (23 cases of TTP (100%), 36 (77%) cases of HUS and 16 (62%) cases of secondary TMA). 54% of HUS needed dialysis compared with 27% of secondary TMA and none of the TTP. 78% of children and in 57% of adults had complete renal function recovery. Mortality was higher in adults (50% in secondary TMA and 17% in TTP). Higher kidney function recovery rates were associated with younger age (OR: 0.947, P 0.04), TTP diagnosis (OR 8.576, p 0.014) and treatment with plasma exchange (OR 4.696 p 0.046).

BASAL CHARACTERISTICS

	ADULTS (n = 63)	CHILDREN (n = 33)
Median age at diagnosis (years)	43,1 ± 16	3,7 ± 3,8
Type of thrombotic microangiopathy		
TTP	23 (36.5%)	0
UHS	14 (22.2%)	33 (100%)
Secondary TMA	26 (41.26%)	0

FACTORS IMPLICATED IN RENAL FUNCTION RECOVERY IN ADULTS PATIENTS

	OR (IC 95%)	p
Age	0.947 (0,912-0,983)	0,004
TTP diagnosis	8,576(1,54-47,75)	0,014
Plasma exchange treatment	4,696(1,027-21,472)	0,046

ADULTS PRESENTATION

	UHS (n=14)	TTP (n=23)	Secondary TMA(n=26)	p
Renal impairment	14 (100%)	16 (69%)	24 (92%)	0,017
Trombocytopenia	10 (71%)	23 (100%)	24 (92%)	0,005
Oliguria	8 (57%)	1 (4%)	10 (38%)	0,002
Diarrhea	7 (50%)	5 (21%)	2 (8%)	0,030
Petechiae	2 (14%)	12 (52%)	4 (15%)	0,011
Plasma exchange	11 (77%)	23 (100%)	16 (62%)	0,005
Dialysis requirement	7 (50%)	0	7 (27%)	0,002
Renal function recovery	8 (57%)	17 (74%)	7 (33%)	0,028
Mortality	0	4 (17%)	13 (50%)	0,001

HEMOLITIC UREMIC SYNDROME

	ADULTS (n=14)	CHILDREN (n=33)	
Age	35,5 ± 11	3,7 ± 3,8	
Anemia	14 (100%)	33 (100%)	
Diarrhea	10 (71%)	10 (30%)	
Shiga toxin test	2/14 (14.3%)	15/33 (45.5%)	
Thrombocytopenia	14 (100%)	25 (75%)	p = 0,007

Pronostic and treatment	ADULTS (n=14)	CHILDREN (n=33)	
Plasma exchange	10 (71%)	3 (9%)	p < 0,001
Dialisys requirement	7 (50%)	25 (75%)	ns
Renal function recovery	8 (57%)	26 (78%)	ns
Mortality	0	2 (6%)	ns

CONCLUSIONS

TMA represents a spectrum of disease consisting of different etiologies associated with very high morbidity and mortality rates. The clinical presentation of TMA guides the diagnosis, but is not enough to establish its etiopathogeny. Therefore, identifying the specific cause for TMA is essential for more accurate diagnosis, more effective treatment, and to predict patient and renal damage prognosis.

1. James N et al. Syndromes of Thrombotic Microangiopathy. N Engl J Med 2014; 371:654-666.
2. Yu-Min She et al. Clinical evaluation of thrombotic microangiopathy: identification of patients with suspected atypical hemolytic uremic syndrome. *Thrombosis Journal*201614(Suppl 1):19.
3. Zayd L et al. Drug-induced thrombotic microangiopathy: a systematic review of published reports. Blood 2015 125:616-618.

DOI: 10.3252/pso.eu.54ERA.2017