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Outcome of Thrombotic Thrombocytopenic Purpura Patients: Single Center Experience

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To the Editor

Thrombotic thrombocytopenic purpura (TTP) is a rare, life-threatening condition.[1,2] It is characterized by platelet-rich thrombi in the microcirculation caused by severely decreased activity of the von Willebrand factor-cleaving protease ADAMTS13 (a disintegrin and metalloprotease with thrombospondin type motif 13), leading to the accumulation of ultra large von Willebrand factor multimers, microangiopathic hemolytic anemia and sometimes organ damage. TTP can be acquired due to an autoantibody inhibitor development which is against to *ADAMTS-13*, or hereditary due to inherited mutations in *ADAMTS13*. Hereditary TTP represents less than 5 percent of all TTP cases; over 95 percent are acquired autoimmune TTP.[3] TTP is a hematologic emergency that is almost always fatal if appropriate treatment is not initiated promptly, even with treatment, the mortality rounds 10% to 20%.[1]

In this retrospective study we aimed to investigate the factors affecting the outcome of TTP patients. Nineteen TTP patients (11 females and 8 males) had a mean age of 41.5±12.7 (18-60) years; 12(63.1%) had neurologic features, 4(21.1%) fever, 3(15.7%) renal impairment (Table-1). All patients received plasma exchange (PEX) therapy within 5 hours of admission. 18 (94.7%) patients received adjunctive 1 mg/kg methylprednisolone (except one hereditary TTP patient). One refractory patient and two relapsed patients received rituximab. Statistical analyses were performed by Jamovi version 0.9.2.6 software. We used the Kruskal Wallis and the Mann-Whitney U test to examine the mean differences. A *P* value of <0.05 was considered as statistically significant.

Laboratory results were presented in Table-2. Relapse/refractory patients and non-relapse/refractory patients were compared in terms of number of PEX sessions until obtaining remission, laboratory values and ADAMTS13 panel.

In conclusion, three interesting results occurred after analysis of data in our study. Firstly, our overall mortality rate was 1 of 19 (5.3%). Higher mortality rates were reported in previous studies (10-20%)[1,2].This result may show that early PEX initiation is an effective factor in mortality reduction. Secondly, the mean d-dimer value of our TTP patients was higher than the reference limit with 2.65 mcg/mL (Reference values 0-0.4 mcg/mL). So, slightly elevated d-dimer level should not hesitate to start urgent PEX treatment in patients with clinically high suspicion of TTP if ADAMTS13 panel results are not obtained quickly. And thirdly, relapsed/refractory patients needed more PEX sessions to achieving first remission. A small number of PEX to achieve response may be predictive of durable remission without relapse.

References

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Characteristics	n	%
Age, mean (range) 41.5 (18-60)		
Gender		
Female	11	
Male	8	
Neurological features		
Syncope	3	15.8
Coma	3	15.8
Confusion	2	10.5
Headache	1	5.3
Dizzness	1	5.3
Seizures	1	5.3
Slurred speech	1	5.3
Renal manifestations	3	15.7
Fever	4	21.1

Parameter	Patients' Mean±SD (min-max) Values	Normal Reference Range
WBC count	8.14±3.37(2.92-16)x10^9/L	4-10x10^9/L
Hemoglobin	8.59±1.63 (5.93-12.4) g/dL	12-16 g/dL
Platelet count	26.42±25.11 (1-110) x10^9/L	150-400x10^9/L
Urea	47.4±19.8 (22-92) mg/dL	15-44 mg/dL
Creatinine	0.98±0.41 (0.5-2.26) mg/dL	0.72-1.25 mg/dL
Total Bilirubin	3.33 (0.72-9.43) mg/dL	0.2-1.2 mg/dL
Indirect Bilirubin	2.42 (0.44-8.89) mg/dL	0.1-0.7 mg/dL
Lactate Dehydrogenase	1186 (228-2570) U/L	125-220 U/L
Alanine Aminotransferase (ALT)	31.2 (10-90)	0-55 U/L
Aspartate Aminotransferase (AST)	41.8 (14-79)	5-34 U/L
Prothrombin Time (PT)	15.4 (12.1-29.4) sn	
INR	1.24±0.39 (0.89-2.75)	1-1.5
Activated Partial thromboplastin time (aPTT)	38.3 (16.1-180) sn	26.5-40 sn
D-Dimer	2.65 (0.54-10.7) mcg/mL	0-0.4 mcg/mL
Fibrinogen	341±106 (149-566) mg/dL	200-400 mg/dL
C-Reactive Protein	20.3±24.1 (0.1-77.8) mg/L	0.1-5 mg/L
Mean platelet volume	10.7±2.85 (5.98-16.2) fL	7.8-11 fL
Mean ADAMTS13 antigen	0.151 (0.02-0.70) mcg/mL	0.60-1.60
Mean ADAMTS13 activity	1.09 (0-8) %	40-130 %
Mean ADAMTS13 inhibitor	45.9 (4.4-90) U/mL	< 12