Original Research Paper

Volume - 13 | Issue - 05 | May - 2023 | PRINT ISSN No. 2249 - 555X | DOI : 10.36106/ijar

General Medicine

OUTCOME IN THROMBOTIC MICROANGIOPATHY WITHOUT ANY OBVIOUS SECONDARY CAUSE

Dr Dnyaneshwar Bonde — Department of Medicine, TNMC and BYL Nair Ch Hospital, Mumbai – 400008

Dr Md Abdurrab Shahbaz Khan* — Medicine department, TNMC and BYL Nair Ch Hospital , Mumbai –400008 .*Corresponding Author

Dr Harshalkumar Jadav — Medicine department, TNMC and BYL Nair Ch Hospital , Mumbai – 400008

KEYWORDS :

INTRODUCTION

Thrombotic microangiopathies are a group of disorders characterized by microangiopathic hemolytic anemia, thrombocytopenia and microthrombi leading to ischemic organ injury. Thrombotic microangiopathies are life-threatening conditions that require urgent management. Thrombocytopenia and microangiopathic hemolytic anemia (MAHA) are defining features of TMAs. Thrombotic microangiopathy has guarded prognosis, more so, when without any obvious secondary cause. Clinical profile & outcome of this group has been studied with focus on renal outcome. Clinical demography & outcome in thrombotic Microangiopathy without any obvious secondary cause

AIMS AND OBJECTIVES

Aim: To study clinical demographic & outcome in thrombotic microangiopathy without any obvious secondarycause. Objective :To study and evaluate the cases of renal dysfunction to look for thrombotic Microangiopathy as a cause and its management.

METHODS AND MATERIALS

This observational study included all patients from Jan2021-July2022. We studied total 1734 cases of renal dysfunction and noted 14 cases having thrombotic microangiopathy. We Studied Clinical demography details. For the diagnosis of TMA, Markers of microangiopathic hemolysis (schistocytes on peripheral smear, thrombocytopenia, raised LDH,l ow serum heptaglobulin) Kidney biopsies, anti factor-HAb, C3NeF were done in all. Genetic studies done wherever financially feasible. TMA diagnosis made with MAHA &/or histopathology. Immunosuppresion started as indicated.Outcome & prognostic factors evaluated.

RESULT

Parameters
MALE:FEMALE 5:9
Age 18-40yrs 11
Age 40-70yrs 3
AKI 11
CKD 3
HYPERTENSION 9
PREGNANCY RELATED 7

SYMPTOMS

uremic symptoms
oliguria /anuria
dyspnoea
edema
fever
headache
diarrhoea

INVESTIGATIONS

- Sr.creatinine M-3.3(1.5-7.6mg/dl). Hb Mean-7.75(4.2-12 gm/dl), Platelete M-128214,(49000-282000)

KIDNEY BIOPSY 11
CT (REVERSE RIM SIGN)- 1 ANTI FACTOR H ANTIBODIES- 5
CFH MUTATION -3 CD 46 MUTATION -1
UNKNOWN CAUSE- 5 C3 NEHRITIC FACTOR 1

TREATMENT

Treatment	No.of patients
HD	11
PLASMAPHERESIS with FFP	11
STEROIDS	8
MMF	4

COMPLICATIONS Steroid induced DM-1, Pulmonary koch's -2.

	COMPLETELY RECOVERED	PARTIALY RECOVERED	HD DEPENDENT	DEATH
TOTAL	5	3	6	3
HD	3	2	6	3
PLEX	5	2	3	2

PREGNANCY RELATED

We have 7 pregnancy related TMA cases 6 were full term ,1 preterm ,LSCS done in 4 ,induced delivery in others 1 IUFD in 7 month ,but no maternal mortality 6 were on PLEX and 7 on HD at presentation 4 completely recovered ,1 partially recovered,2 were HD dependent of which 1 kidney transplantation done after 6 month, but had graft failure

CONCLUSION

Early detection, prompt treatment in the form of plasmapheresis and immunosuppression may prevent patient from progressing to ESRD.

REFERENCES

1. Brocklebank V, Wood KM, Kavanagh D. Thrombotic microangiopathy and the kidney. Clin J Am Soc Nephrol . 2018;13(2):300–317. 2. Goodship TH, Cook HT, Fakhouri F, et al.
2. Atypical hemolytic uremic syndrome and C3 glomerulopathy: conclusions from a “Kidney Disease: Improving Global Outcomes” (KDIGO) Controversies Conference. Kidney Int. 2017;91(3):539–551. 3. Tarr PI, Gordon CA, Chandler WL.
3. Shiga-toxin-producing Escherichia coli and hemolytic uremic syndrome. Lancet. 2005;365(9464): 1073–1086. 4. Blood . 2008;112(13):4948–4952.Kavanagh D, Richards A, Goodship T, et al.
4. Transplantation in atypical hemolytic uremic syndrome. Semin Thromb Hemost . 2010;36(6):653–659. 5. Bruel A, Kavanagh D, Noris M, et al.
5. Hemolytic uremic syndrome in pregnancy and postpartum. Clin J Am Soc Nephrol . 2017;12(8): 1237–1247.
6. George JN, Nester CM, McIntosh JJ. Syndromes of thrombotic microangiopathy associated with pregnancy. Hematology Am Soc Hematol Educ Program . 2015;2015:644–648.