Severe form of fulminant hepatic failure (Wilson's disease) and hepatorenal syndrome treated with combination of CRRT and TPE leading to successful liver transplantation

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Introduction

Wilson's disease is an inherited metabolic disorder that results in copper accumulation mainly in liver and brain. In rare occasions, the disease can present with fulminant liver failure (FHF), a sudden deterioration of liver function in an otherwise healthy individual associated with : a) jaundice, b) coagulopathy, c) any degree of encephalopathy and d)multiple organ dysfunction (50% renal insufficiency).

Case description

An eight-year-old boy was admitted to PICU due to fulminant hepatic failure. Clinical deterioration was acute at 1st day of hospitalization in PICU with acute kidney injury (hepatorenal syndrome), mental deterioration and massive intestinal hemorrhage. Hemorrhage was controlled. Because of the severity of his condition, he was placed on mechanical ventilation, invasive monitoring of intracranial pressure and continuous renal replacement treatment (CVVH). Laboratory tests as well as clinical approach suggested Wilson's disease. In order to facilitate the urinary excretion of copper, D-penicillamine was used. The patient was placed in high priority on the transplant list. Though seeking medical interventions to provide time until finding liver donor, extraction of circulating copper by plasmapheresis was suggested. He underwent treatment of plasmapheresis daily without complications. After 9 days of hospitalization, he was transferred to the liver transplantation center.

Table 1.Laboratory findings

	Before admission	Day 1	Day 2	Day 3	Day 3 *	Day 5	Day 8
Hbg	4.4	6.6	8.1	7.7	6.2	9.9	9.1
WBC	24,500	16,540	10,330	10,680	8,620	11,310	13,640
TBIL (S)	42.6	45.4	50.1	51.8	44.7	35.6	35
DBIL (S)	32	35.6	49.5	38.7	18.7	29.6	30
AST	171	197	216	214	98	186	125
ALT	10	14	34	26	18	80	73
γ- GT	145	127	78	63	52	31	38
INR	5.1	5.1	3.2	3.75	1.87	2.01	2.17
UREA	137	147	188	200	215	85	147
CREA	1.9	1.6	2.7	3	4.1	1.6	2
COPPER (U)		1386					
Na	135	135	137.7	142.9	156	146	143
K	4.5	3.4	3.3	3.2	2.7	3.5	3.3
LDH	978	1,225	1,643		592	717	535

* After 1st TPE

Conclusion

Plasmapheresis combined with CRRT could be a successful medical treatment in fulminant Wilson disease and should be considered as a therapeutic measure to stabilize a patient by decreasing serum copper, reducing hemolysis and maintaining homeostasis.

References

1. Ala, A., et al., *Wilson's disease*. Lancet, 2007. **369**(9559): p. 397-408.4
2. Roberts, E.A., M.L. Schilsky, and D. American Association for Study of Liver, *Diagnosis and treatment of Wilson disease: an update*. Hepatology, 2008. **47**(6): p. 2089-111.
3. Walshe, J.M., *Penicillamine: the treatment of first choice for patients with Wilson's disease*. Mov Disord, 1999. **14**(4): p. 545-50.
4. Das, S.K. and K. Ray, *Wilson's disease: an update*. Nat Clin Pract Neurol, 2006. **2**(9): p. 482-93.