

Table 1: Demographic Characteristics of the Patients with TAFRO Syndrome and idiopathic Multicentric Castleman Disease

Median value (range)	TAFRO (n = 8)	iMCD (n = 7)	P Value
Age	43.5 (19-81)	48 (38-73)	0.6025
Male (%)	5 (62.5)	3 (42.3)	0.6193
WBC ($10^9/L$)	13.4 (4.6-18.3)	6.9 (4.0-20.6)	0.0933
Neutrophils (%)	83.7 (77-93)	69.1 (59.0-81.0)	<0.01**
Hemoglobin (g/L)	117 (82-138)	104 (69-149)	0.4871
Platelet ($10^9/\mu L$)	38.5 (8.0-91.0)	354 (223-595)	<0.01**
D-dimer ($\mu g/L$)	19.8 (3.3-48)	0.8 (0.5-34.9)	0.0922
BUN (mg/dL)	30.7 (6.2-92.9)	14.5 (10.7-20.9)	0.2972
Creatinine ($\mu mol/L$)	132.6 (39.8-219.2)	59.2 (46.9-171.5)	0.0728
Total protein (g/L)	47 (38-61)	96 (72-115)	<0.01**
Albumin (g/L)	19 (11-23)	24 (21-35)	<0.01**
Total Bilirubin ($\mu mol/L$)	17.6 (5.5-151.0)	8.6 (4.8-12.7)	<0.01**
ALP (U/L)	550 (195-1357)	238 (116-436)	0.1182
γ -GTP (U/L)	78 (11-166)	28 (7-61)	0.0562
Cholinesterase (U/L)	58 (32-80)	146 (103-347)	<0.01**
Soluble IL-2 receptor (U/mL)	1975 (660-3831)	1382 (856-5376)	0.7723
Ferritin ($\mu g/L$)	577.6 (386.3-994.5)	192.4 (17.4-303.9)	<0.01**
IgG (g/L)	9.9 (8.5-22.1)	41.0 (28.3-67.2)	<0.01**
IgG4 (g/L)	0.20 (0.030-0.30)	3.1 (1.1-15.0)	<0.01**
Erythrocyte sedimentation rate (mm/h)	79 (46-129)	119 (83-140)	<0.05*
C-reactive protein (mg/L)	178 (72.3-282.0)	66.9 (20.5-126.0)	<0.01**
Procalcitonin (ng/mL)	4.76 (0.378-10.8)	0.093 (0.020-0.576)	<0.01**
Interleukin-6 (pg/mL)	28.0 (17.2-141)	23.3 (8.6-71.6)	0.2013
VEGF (pg/mL)	684 (215-1710)	1091 (256-1880)	0.4555

Statistical significance was calculated using Wilcoxon / Kruskal-Wallis test. * $p < 0.05$,

** $p < 0.01$, *** $p < 0.001$.

Table 2: Clinical Characteristics of the Patients with TAFRO Syndrome and idiopathic Multicentric Castleman Disease

Median value (range)	TAFRO (n = 8)	iMCD (n = 7)	P Value
Days Until Diagnosis	10 (5-47)	13 (11-62)	0.1865
Length of Hospitalization (Day)	87 (34-236)	30 (13-59)	<0.01**
First Chief Complaints			
Fever, n (%)	5 (62.5)	3 (42.9)	
Abdominal Pain, n (%)	4 (50.0)	2 (28.6)	
Cough, n (%)	2 (25.0)	0	
Generalized Fatigue, n (%)	0	2 (28.6)	
Shortness of Breath, n (%)	1 (12.5)	2 (28.6)	
First Diagnosis			
Infectious Disease, n (%)	6 (75.0)	1 (14.3)	
Autoimmune Disease, n (%)	1 (12.5)	2 (28.6)	
Malignancy, n (%)	0	2 (28.6)	
IgG4-RD, n (%)	0	1 (14.3)	
Small Bowel Obstruction, n (%)	0	1 (14.3)	
Heart Failure, n (%)	1 (12.5)	0	
Reasons for Hospitalization			
Fever of Unknown Origin, n (%)	4 (50.0)	0	
Abdominal Pain, n (%)	3 (37.5)	2 (28.6)	
Acute Kidney Injury, n (%)	1 (12.5)	0	
Pleural Effusion, n (%)	0	2 (28.6)	
Suspected Infection, n (%)	0	1 (14.3)	
Generalized Fatigue, n (%)	0	1 (14.3)	
Anemia, n (%)	0	1 (14.3)	
Symptoms			
Fever, n (%)	7 (87.5)	2 (28.6)	<0.05*
Fever type			
Remittent, n (%)	6 (85.7)	0	0.0833
Intermittent, n (%)	1 (14.3)	2 (100)	
Night Sweat, n (%)	0	1 (14.3)	0.4667
Weight Loss, n (%)	0	1 (14.3)	0.4667

Lymphadenopathy, n (%)	5 (62.5)	7 (100)	0.2000
Organomegaly, n (%)	9 (100)	7 (100)	0
Pleural effusion / Ascites, n (%)	7 (87.5)	2 (28.6)	<0.05*
Lower extremity edema, n (%)	7 (87.5)	1 (14.3)	<0.05*
Treatments			
1 st line			
Methylprednisolone pulse, n (%)	6 (75.0)	0	
Prednisolone 1mg/kg, n (%)	2 (25.0)	2 (28.6)	
Prednisolone ≤ 0.5mg/kg, n (%)	0	5 (71.4)	
2 nd line			
Tocilizumab, n (%)	2 (25.0)	3 (42.8)	
Rituximab, n (%)	3 (37.5)	0	
Cyclosporine, n (%)	2 (25.0)	0	
Etoposide, n (%)	0	1 (14.3)	
Melphalan, n (%)	0	1 (14.3)	
Bortezomib, n (%)	1 (12.5)	0	
No 2 nd line treatment, n (%)	0	2 (28.6)	
3 rd line			
Tocilizumab, n (%)	3 (50.0)	1 (50.0)	
Rituximab, n (%)	2 (33.3)	1 (50.0)	
Cyclophosphamide + Vincristine, n (%)	1 (16.7)	0	
Death, n (%)	1 (12.5)	1 (14.3)	

Statistical significance was calculated using Wilcoxon / Kruskal-Wallis test. The Fisher's

exact analysis was used for the statistical analysis of nominal scales. * $p < 0.05$, ** $p < 0.01$, *** $p < 0.001$.