

Supplementary Table. Patients characteristics, treatment and outcomes.

Patient	Leading clinical symptoms/signs at first referral	Basic laboratory results at admission to hospital	ADAMTS-13 activity	Functional complement analysis	Immunoserological analysis	Basic laboratory results on discharge from hospital and/or outpatient visit	Outcome
Pt#1 F, 35y	Headache, spontaneous mucosal bleeding, suffusions, severe thrombocytopenia.	15.09.2022: Lkci 4,2 10 <sup>9</sup> /L; Erci 3,08 10 <sup>12</sup> /L; Hb 82 g/L; Tr 37 10 <sup>9</sup> /L; BUN 3,4 mmol/l; Creatinine 79 µmol/l; eGF (CKD-EPI)/1,73 m2 84 ml/min; Bilirubin tot. 28 µmol/l; Bilirubin dir. 10 µmol/l; Albumin 46 g/l; LDH 7,41 µkat/l; Shistocytes 7,8%.	1,7 %.	CP activity (haemolitical test): 144 %; AP activity (ELISA AP): 111 %; LP activity: 104 %; Complement component factor B: 425 mg/l; factor H: 514 mg/l; factor I: 32 mg/l; C3: 1,09 g/l; C4: 0,28 g/l; SC5b-9 litic complex: 141 ng/ml; Urinary SC5b-9 litic complex: 50 ng/ml.	ANA negative; anti-ENA- Sm,U1RNP,Ro,La,Scl-70,Jo-1,PCNA,PM/ScI,SL,Ku negative; aCL IgG <5, IgM <5; anti-beta2GPI IgG <2, IgM <2, IgA <2; aPS/PT IgG <5, IgM <5, IgA <5.	20.09.2022: Lkci 9,5 10 <sup>9</sup> /L; Erci 3,08 10 <sup>12</sup> /L; K-Hb 87 g/L; Tr 249 10 <sup>9</sup> /L; BUN 5,5 mmol/l; Creatinine 77 µmol/l; eGF (CKD-EPI)/1,73 m2 86 ml/min; Bilirubin tot. 7 µmol/l; Bilirubin dir. 3 µmol/l; Albumin 41 g/l; LDH 2,38 µkat/l.	Complete resolution of symptoms/signs of the disease.
Pt#2 M, 43y	Malignant hypertension (RR>230/125 mmHg), retinal haemorrhages, acute kidney injury.	23.02.2021: Lkci 10,0 10 <sup>9</sup> /L; Erci 3,48 10 <sup>12</sup> /L; K-Hb 108 g/L; Tr 106 10 <sup>9</sup> /L; BUN 26,4 mmol/l; Creatinine 536 µmol/l; eGF (CKD-EPI)/1,73 m2 10 ml/min; Bilirubin tot. 23 µmol/l; Bilirubin dir. 6 µmol/l; Albumin g/l 41; LDH 5,18 µkat/l.	45,1 %.	CP activity (haemolitical test) 103 %; AP activity (ELISA AP): 104 %; LP activity 170 %; Complement component factor B: 402 mg/l; factor H: 376 mg/l; factor I: 35 mg/L; C3: 0,90 g/l; C4: 0,29 g/l; SC5b-9 litic complex: 236 ng/ml; C3Nef: 5,1% - negative; Antibodies against factor H (anti-FH): 2,6 U/ml – negative, Antibodies against C1q (anti-C1q): 2,7 U/ml - negative.	anti-ds DNA negative; ANA positive, titer 1:640; anti-ENA- Sm,U1RNP,Ro,La,Scl-70,Jo-1,PCNA,PM/ScI,SL,Ku negative; anti-CCP <25; aCL IgG 6, IgM <5, IgA <5; anti-beta2GPI IgG <2, IgM <2, IgA <5.	6.4.2021: Lkci 10,8 10 <sup>9</sup> /L; Erci 4,51 10 <sup>12</sup> /L; K-Hb 139 g/L; Tr 249 10 <sup>9</sup> /L; BUN 23,1 mmol/l; Creatinine 727 µmol/l*; eGF (CKD-EPI)/1,73 m2 7 ml/min*; Bilirubin tot. 6 µmol/l; Bilirubin dir. 2 µmol/l; Albumin 46 g/l; LDH 3,44 µkat/l. *dialysis dependant	Resolution of symptoms/signs of malignant hypertension, ESRD. Kidney transplant (deceased donor) July 2022, no TMA recurrence.

Pt#3 M, 32y	Nausea, vomiting, oliguric acute kidney injury.	29.09.2021 Lkci 4,6 10^9/L; Erci 2,79 10^12/L; K-Hb 86 g/L; Tr 120 10^9/L; BUN 19,2 mmol/l; Creatinine 214 μmol/l; eGF (CKD- EPI)/1,73 m2 34 ml/min; Bilirubin tot. 27 μmol/l; Bilirubin dir. 9 μmol/l; Albumin 35 g/l; LDH 7,74 μkat/l.	74,8 %.	CP activity (haemolytic test): 102 %; ( ANA negative; AP activity (ELISA AP): 82 %; anti-ENA - LP activity: 13 %; complement Sm,U1RNP,Ro,La,Scl-70,Jo- component 1,PCNA,PM/Scl,SL,Ku factor B: 277 mg/l; factor H: 378 mg/l; factor I: 24 mg/l; C3: 0,7 g/l; C4: 0,14 g/l; SC5b-9 litic complex: 198 ng/ml; Urinary C5b-9 litic complex: 8 ng/ml.	4.10.2021 Lkci 3,97 10^9 /L; Hb 73 g/L;Tr 282 10^9 /L; BUN 7,1 mmol/l; Creatinine 126 μmol/l; eGF (CKD-EPI)/1,73 m2 65 ml/min; LDH 5,02 μkat/l.	Complete resolution of symptoms/signs of the disease.	
		29.10.2021: Lkci 4,7 10^9/L; Erci 3,90 10^12/L; Hb 122 g/L; Tr 173 10^9/L; BUN 3,7 mmol/L; Creatinine 96 μmol/L; eGF (CKD-EPI)/1,73 m2 90 mL/min; Bilirubin tot. 15 μmol/L; Bilirubin dir. 4 μmol/L; Albumin 46 g/L.					
Pt#4 M, 33y	Malignant hypertension (RR 258/148 mmHg), blindness (retinal haemorrhages and edema of the optic nerve papilla), acute kidney injury, nausea, vomiting, severe thrombocytopenia, hemolytic anaemia.	14.10.2021 Lkci 6,7 10^9/L; Erci 2,10 10^12/L; Hb 66 g/l; Tr 29 10^9/L; BUN 46,2 mmol/l; Creatinine 819 μmol/l; eGF (CKD- EPI)/1,73 m2 7 ml/min; Bilirubin tot. 23 μmol/l; Bilirubin dir. 7 μmol/l; Albumin 36 g/l; LDH 26,70 μkat/l; Shistocytes 6,7%.	57,1 %.	CP activity (haemolytic test): 127 %; AP activity (ELISA AP): 115 %; LP activity 25 %; Complement component factor B: 314 mg/l; factor H: 439 mg/l; factor I: 29 mg/l; C3: 0,71 g/l; C4: 0,22 g/l; SC5b-9 litic complex 182 ng/ml; Urinary C5b-9 litic complex >181 ng/ml; C3Nef: 0 % - negative; Antibodies against factor H (anti-FH): 0,7 U/ml – negative; Antibodies against C1q (anti- C1q): 1,3 U/ml – negative.	ANA negative; anti-ENA- Sm,U1RNP,Ro,La,Scl-70,Jo- 1,PCNA,PM/Scl,SL,Ku negative; aCL IgG <5, IgM <5; anti-beta2GPI IgG <2, IgM <2, IgA <2; aPS/PT IgG <5, IgM <5, IgA <5; ANCA negative.	15.11.2021 Lkci 5,6 10^9/L; Erci 3,70 10^12/L; Hb 119 g/l; Tr 12 10^9/L; BUN 28,1; Creatinine 806 μmol/l*; eGF (CKD-EPI)/1,73 m2 7 ml/min*; Bilirubin tot. 8 μmol/l; Bilirubin dir 2 μmol/l; Albumin 47 g/l; LDH 3,98 μkat/l. *dialysis dependant	Improvement of blood pressure and hematologic disease, severe visual impairment persists, ESRD.
Pt#5 M, 25y	Nausea, vomiting, diarrhoea, acute kidney injury,	10.1.2022	49,2%.	CP activity (haemolytic test): 114 %; AP activity	ANA negative; anti-ENA- Sm,U1RNP,Ro,La,Scl-70,Jo-	24.02.2022 Lkci 11,28 10^9/L; Hb 108 g/l; Erci 3,51 2,10 10^12/L;	Improvement of gastrointestinal symptoms and hematologic disease,

	thrombocytopenia, hemolytic anaemia.	Lkci 4,0 10 <sup>9</sup> /L; Erci 2,33 2,10 10 <sup>12</sup> /L; Hb 72 g/L; Tr 58 10 <sup>9</sup> /L; BUN 47,3 mmol/L; Creatinine 1313 µmol/L; eGF (CKD-EPI)/1,73 m2 4 ml/min; Bilirubin tot. 5 µmol/L; Bilirubin dir. 2 µmol/L; Albumin 33 g/L; LDH 7,14 µkat/L; Shistocytes 4%.		(ELISA AP): 112 %; LP activity: 125 %; complement component factor B: 457 mg/L; factor H: 436 mg/L; factor I: 42 mg/L; C3: 0,93 g/L; C4: 0,23 g/L; C5b-9 litic complex: 350 ng/ml; C3Nef: 3,6 %-negative; Antibodies against factor H (anti-FH): 2,8 U/ml-negative; Antibodies against C1q (anti-C1q): 5,2 U/ml-negative.	1,PCNA,PM/ScI,SL,Ku negative; aCL IgG <5, IgM <5; anti-beta2GPI IgG <2, IgM <2, IgA <2; aPS/PT IgG <5, IgM <5, IgA <5; ANCA negative.	Tr 121 10 <sup>9</sup> /L; BUN 27,2 mmol/L; Creatinine 679 µmol/L *; eGF (CKD-EPI)/1,73 m2 9 ml/min*; Bilirubin tot. 7 µmol/L; Bilirubin dir. 3 µmol/L; Albumin 35 g/L; LDH 5,48 µkat/L. *dialysis dependant	ESRD. Kidney transplant (deceased donor) April 2024- no TMA recurrence.
Pt#6 M, 52y	Nausea, propulsive vomiting, severe worsening of chronic kidney transplant disease* (uremia) *pt with transplanted kidney, creatinine at last follow up before referral was 181 µmol/L, eGF 36 ml/min.	26.2.2021 Lkci 5,4 10 <sup>9</sup> /L; Erci 2,82 10 <sup>12</sup> /L; Hb 80 g/L; Tr 179 10 <sup>9</sup> /L; BUN 15,2 mmol/L; Creatinine 688 µmol/L*; Bilirubin tot. 10 µmol/L; Bilirubin dir. 6 µmol/L; Albumin 30 g/L; LDH 3,24 µkat/L; Tacrolimus 4,5 µg/L. ** hemodialysis performed in the emergency room before admission due to uremia (Creatinine>2000 µmol/L)	Not performed.	Not performed.	anti-CCP <25; aCL IgG <5, IgM <5; IgA <5; anti-beta2GPI IgG <2, IgM <2, IgA <2; aPS/PT IgG <5, IgM <5, IgA <5.	26.4.2021 Lkci 6,1 10 <sup>9</sup> /L; Erci 3,11 2,82 10 <sup>12</sup> /L; K-Hb 98 g/L; Tr 154 10 <sup>9</sup> /L; Tacrolimus 6,5 µg/L; BUN 25,2 mmol/L; Creatinine 520 µmol/L; eGF (CKD-EPI)/1,73 m2 10 ml/min; Bilirubin tot. 13 µmol/L; Bilirubin dir. 5 µmol/L; Albumin 34 g/L; LDH 4,09 µkat/L.	Partial recovery of kidney graft function - at the time of discharge he was not on dialysis. Two months later ESRD, start of dialysis. 2nd Kidney transplant (deceased donor) August 2024, no TMA recurrence.
Pt#7 M, 33y	Fever, cough, diarrhoea, signs of malignant hypertension (RR 210/110), visual	30.8.2024 Lkci 11,7 10 <sup>9</sup> /L; Erci 2,81 10 <sup>12</sup> /L; Hb 87 g/L; Tr 78 10 <sup>9</sup> /L; BUN 32,2 mmol/L; Creatinine 1072	80,1 %.	CP activity (haemolytic test): 94 %; AP activity (ELISA AP): 96 %; LP activity: 27 %; Complement component factor B: 238 mg/L;	RF <15,9 IU/mL-negative; ANCA negative; Anti-GBM negative; Anti-C1q negative; Anti-PLA2R IgG negative; Anti-THSD7A IgG negative;	13.9.2024 Lkci 12,7 10 <sup>9</sup> /L; Erci 2,82 10 <sup>12</sup> /L; Hb 88 g/L; Tr 177 10 <sup>9</sup> /L; BUN 27,8 mmol/L; Creatinine 803 µmol/L *;	Resolution of symptoms/signs of malignant hypertension, ESRD.

	disturbances, acute kidney injury.	μmol/L; eGF (CKD-EPI)/1,73 m2 <b>5</b> mL/min; Bilirubin tot. 10 μmol/L; Bilirubin dir. 4 μmol/L; LDH <b>8,15</b> μkat/L; Haptoglobin <b>&lt;0,07</b> g/l.		factor H: <b>677</b> mg/l; factor I: 35 mg/l; C3: 0,74 g/l; C4: 0,27 g/l; SC5b-9 litic complex: 121 ng/ml; Bb: <b>3,03</b> μg/ml; antibodies against factor H (anti-FH): 1,75 U/ml -negative; C3Nef: 4,02 % -negative; Antibodies against C1q (anti-C1q): 1,18 U/ml-negative.	ANA negative; anti ENA-Sm,U1RNP,Ro,La,Scl-70,Jo-1,PCNA,PM/Scl,SL,Ku negative; aCL IgG <5, IgM <5; IgA <5; anti-beta2GPI IgG <2, IgM <2, IgA <2; aPS/PT IgG <5, IgM <5, IgA <5; lupus anticoagulants-negative.	eGF (CKD-EPI)/1,73 m2 <b>7</b> mL/min*; Bilirubin tot. 5 μmol/L; Bilirubin dir. 2 μmol/L; Albumin 38 g/l; LDH 3,32 μkat/l. *dialysis dependant	
<b>Pt#8 F, 29y</b>		13.8.2024 Lkci 4,2 10^9/L; Erci 2,75 10^12/L; K-Hb <b>83</b> g/l; Tr 152 10^9/L; BUN <b>27,7</b> mmol/L; Creatinine <b>720</b> μmol/L; eGF (CKD-EPI)/1,73 m2 <b>6</b> mL/min; Bilirubin tot. 12 μmol/L; Bilirubin dir. 3 μmol/L; Albumin 37 g/l; LDH 3,92 μkat/L; Haptoglobin 1,30 g/l, Shistocytes <b>2,1</b> %.		CP activity (haemolytic test): 139 %; AP activity (ELISA AP): 93 %; LP activity: 79 %; complement component factor H: 562 mg/l; factor I: 29 mg/l; factor B: 240 mg/l; C3: 0,94 g/l; C4: 0,22 g/l; SC5b-9 litic complex: 186 ng/ml; Bb: <b>1,95</b> μg/ml.	ANA negative; anti-ENA-Sm,U1RNP,Ro,La,Scl-70,Jo-1,PCNA,PM/Scl,SL,Ku negative; antibodies associated with SSc (Scl-70 <5 AU; CENP-A <5 AU; CENP-B <5 AU; PM-Scl 100 <5 AU; PM-Scl 75 <5 AU; Ku <5 AU; RNA Polimeraza III <5 AU; RNP 68kD/A/C <5 AU; Th/To <5 AU; Fibrilarin <5 AU; NOR-90 <5 AU; SSA/Ro52kD <5 AU)-negative.	23.8.2024 Lkci 7,5 10^9/L;-Erci 2,88 10^12/L; Hb <b>87</b> g/L; Tr 197 10^9/L; Haptoglobin 1,62 g/L; BUN <b>31,6</b> mmol/L; Creatinine <b>771</b> μmol/L; eGF (CKD-EPI)/1,73 m2 <b>6</b> mL/min; LDH 3,58 μkat/L.	
	Fatigue, high blood pressure (180/110 mmHg), acute worsening of chronic kidney disease - at the last follow-up examination she had a Creatinine of 117 μmol/l, eGF 55 ml/min.	Not performed				Improvement of high blood pressure, ESRD. On hemodialysis since 6.9.2024.	

**Legend of abbreviations and reference values of selected laboratory parameters (significant deviations in laboratory values are highlighted):**

ANA-antinuclear antibodies, ENA-extractable antibody nuclear panel, aCl-anticardiolipin antibodies, anti-beta2GPI- Beta 2 glycoprotein I antibodies, aPS/PT-antithrombin antibodies, ANCA- anti neutrophil cytoplasm antibodies,

CP-classical complement pathway (normal value for adults: 50-150%),

AP-alternative complement pathway (normal value for adults: 30 - 113%),

LP-lectin complement pathway (normal value for adults: 0 - 125%),

C3-complement component C3 (reference value for adults 0,60 - 1,30 g/l),

C4-complement component C4 (reference value for adults 0,10 - 0,30 g/l),

C5b-9-membrane attack complex, litic complex (reference value for adults in plasma: 127 - 303 ng/ml; reference value for adults in urin: < 30 ng/ml),

Factor B-(reference value for adults 205-400 mg/l),  
Factor H-(reference value for adults 380,6-674,9 mg/l),  
Factor I-(reference value for adults 21,2-42,1 mg/l),  
Bb- (reference value for adults 0,49 - 1,42 µg/ml),

LDH-lactate dehydrogenase (normal value <4,13 µkat/l),

Bilirubin tot.-total bilirubin (normal value<15 µmol/l),

Bilirubin dir.-direct bilirubin (normal value<5 µmol/l),

Haptoglobin (normal value 0,3-2,0 g/