Myocardial dysfunction in life-threatening idiopathic systemic capillary leak syndrome

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Argomento: Funzione cardiovascolare in terapia intensiva

The Idiopathic Systemic Capillary Leak Syndrome (ISCLS) is a rare and potentially fatal disease of unknown aetiology. Episodically, abrupt increases in microvascular permeability occur due to disruption of endothelial adherens junctions (AJ), resulting in a shift of proteinaceous fluid from the intravascular space into the interstitium. As a result, hypotension and extensive oedema develop, leading to multiple organ dysfunctions. The end-organ damage is mainly induced by hypovolaemia, although a role for oedematous congestion has also been hypothesized. Generalized myocardial oedema has already been reported, albeit it is not clear whether it contributes to shock. Here, we present three cases of life-threatening ISCLS in which myocardial involvement seemed to play a crucial role in the pathophysiology of shock.

Haemodynamic values and blood tests results obtained throughout ICU stay are shown in Figure 1 and Table 1. All patients showed variable degrees of myocardial oedema with impaired ventricular filling (Figure 2). Two patients developed signs of cardiogenic as well as hypovolemic shock and needed sedation, mechanical ventilation plus either inotropic treatment (PT1) or Veno-Arterial Extracorporeal Membrane Oxygenation (PT2). Conversely, the third patient (PT3) had a milder presentation and was managed conservatively, with targeted colloid boluses to revert either systolic pressure drops below 70 mmHg or symptomatic hypotensive episodes. In all patients, the acute episode resolved 2 to 8 days after shock onset, with complete reversal of myocardial oedema.

We hypothesize that myocardial dysfunction due to oedema contributes to the development of shock during life-threatening SCLS crises. The endothelial AJ disruption that affects the systemic circulation might as well involve the endocardium, causing myocardial oedema. The resulting diastolic dysfunction combined with hypovolemia precipitates preload deficiency. We strongly encourage the use of repeated, bedside echocardiographic assessments during ISCLS crises to guide fluid therapy and to rapidly detect the need for escalation of haemodynamic support.

Table 1

Blood samples analysis and fluid balances of the three subjects during the first three days of their ICU stay.

	Case 1			Case 2			Case 3		
	Day 1	Day 2	Day 3	Day 1	Day 2	Day 3	Day 1	Day 2	Day 3
Hb (g/dL)	21.9	20.2	14.2	10.6	8.3	10.2	21.9	18	10.5
Ht (%)	62.7	57.9	39.6	35.3	23.7	29.8	64.5	53.4	30.5
RBC (cells/mm^3)	6750	6350	4450	3570	2790	3470	7920	6600	3840
WBC (cells/mm^3)	51690	49230	28620	30740	18390	7010	59400	42470	13840
Neutrophils (%)	77.2	82.3	80.7	79.1	80	82.2	75.6	80.8	81.9
Lymphocytes (%)	19.1	12.5	15	14.3	15.6	11.2	18.5	11.8	19
Monocytes (%)	3.4	4.7	3.9	5.5	3.6	6.5	5.3	7.5	8.6
Eosinophils (%)	0.3	0.5	0.4	1.1	0.8	0.1	0.2	0.1	1
PLT (plt/mm^3)	273000	186000	98000	106000	50000	28000	231000	246000	123000
PT (INR)	2.92	3.44	2.09	5.68	2.53	0.95	1.31	1.28	1.2
PTT (INR)	*n.c.*	*n.c.*	1.81	1.8	1.27	1.13	1.62	1.27	1.15
Fybrinogen (mg/dL)	148	114	194	278	490	675	262	403	605
D-dimer (μg/L)	2880	586	1129	-	-	747	585	789	388
Urea (mg/dL)	-	71	96	91	144	-	67	102	126
Creatinine (mg/dL)	1.93	4.64	5.42	3.37	5.33	6.09	2.2	3.74	2.1
Glucose (mg/dL)	262	139	121	204	132	112	172	95	97
Na (mEq/L)	135	146	145	143	145	146	134	140	140
K (mEq/L)	4.2	3.8	3.7	4.5	4.6	4.2	5.6	5	3.5
Cl (mEq/L)	112	117	110	113	111	108	111	102	94
Ca (mEq/L)	6.1	6	7.2	8	7.7	3.3	8	8	7.9
P (mg/dL)	-	7.2	-	-	-	2.6	6.8	-	4.1
Mg (mg/dL)	2.7	2.1	1.7	1.7	-	5.5	2.2	1.8	2.1
Albumin (g/L)	9	<9.00	16	1.8	-	-	2.5	3	3.6
γGT (U/L)	43	-	-	15	-	213	15	15	16
AST (U/L)	-	1394	-	170	-	97	17	107	183
ALT (U/L)	-	1154	2767	28	38	7	5	16	31
CPK (U/L)	-	267	> 21335	194	226	79	129	5059	10886
LDH (U/L)	-	2639	> 3325	*hem.*	1296	952	279	529	705
Bilirubin (mg/dL)	< 0.1	0.26	1.14	<1.2	<1.2	<1.2	0.4	0.6	0.6
C-reactive protein (mg/L)	-	20.1	28.6	8.1	88	52.5	7	42	67
Daily fluid balance (ml)	4900	2300	-1000	3500	4200	-3700	1450	-2500	-2650
Cumulative fluid balance (ml)			6200			4000			-3700

n.c., uncoagulable; *hem.*, hemolysis.

Figure 1

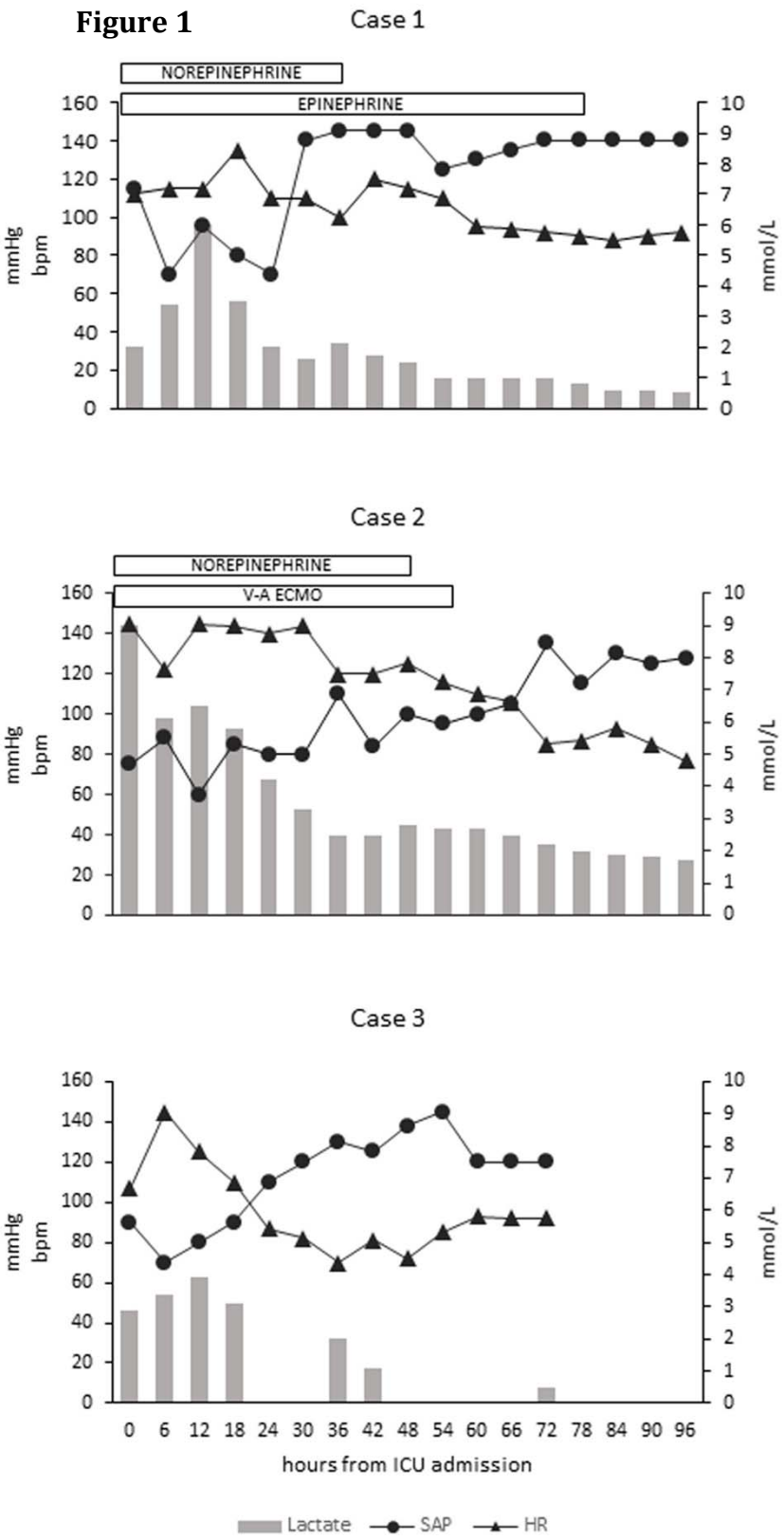

Figure 2