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Case Report

Idiopathic systemic capillary leak syndrome presenting as septic shock: A case report

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ABSTRACT

Background: Idiopathic capillary leak syndrome (Clarkson's Disease) is a rare angiopathy with a heterogeneous phenotype that may present as distributive shock refractory to resuscitative management.**Objective:** We report a case of idiopathic systemic capillary leak syndrome presenting as septic shock.**Methods:** Structured case report and review of the literature.**Results:** A 27-year old man admitted to our institution with coryzal symptoms rapidly deteriorated with presumed sepsis, leading to intensive care unit admission. Following further deterioration, Idiopathic systemic capillary leak syndrome was considered and intravenous immunoglobulin administered, resulting in rapid improvement in the patient's clinical status.**Conclusions:** Idiopathic systemic capillary leak syndrome is a rare and potentially life-threatening angiopathy that may present as, and should be considered in, refractory distributive shock. Administration of intravenous immunoglobulin resulted in rapid recovery in this patient, and has been associated with positive outcomes in previous cases.

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Introduction

Idiopathic systemic capillary leak syndrome is a rare cyclical syndrome characterized by generalized edema, hypotension, hypoalbuminemia and hemoconcentration with visceral sparing.¹ Approximately 168 cases have been reported since Clarkson's original description in 1960,²⁻⁴ but the pathophysiology remains poorly understood. We review the clinical course and management of a patient admitted to our intensive care unit (ICU) with suspected sepsis refractory to resuscitation and vasopressor support, diagnosed with ISCLS. Video of echocardiographic findings is included in the online version of this article.

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Case report

Prodromal phase

A 27-year-old patient presented to the Emergency Department with a ten-day history of fever, myalgia, malaise, a dry cough and widespread macular rash with an increased white cell count (WCC) ($22.78 \times 10^9/L$) and mild decrease in serum albumin concentration (31 g/L, normal range 38 – 40 g/L). Blood and urine specimens were sent for cultures. Viral reverse-transcriptase-polymerase-chain-reaction (RT-PCR) assays, atypical pneumonia serology, urinary legionella and pneumococcal antigen immunochromatography specimens were collected.

Leak phase

On Day 2, the patient reported neck stiffness and photophobia. He was mildly tachycardic (HR 110 bpm), with normal neurological findings; white cell count was $17.22 \times 10^9/L$, lactate concentration was 2.7 mmol/L. Magnesium (0.64 mmol/L) and phosphate

(0.64 mmol/L) levels were decreased. Creatine kinase was elevated at 178 U/L, as was C-reactive protein concentration at 87 mg/L. Albumin and total protein concentrations had fallen to 22 g/L and 40 g/L respectively. Remaining electrolyte levels and renal and liver function tests were otherwise normal. Facial edema was noted. Urgent chest radiography, performed on the ward, was unremarkable. Empiric therapy for meningitis and encephalitis (ceftriaxone, vancomycin, acyclovir and dexamethasone) was commenced in line with Australian national guidelines (Therapeutic Guidelines: Antibiotics).^{5,6} Clinical examination revealed no evidence of raised intracranial pressure and computed tomography scan showed no contra-indications to lumbar puncture. The patient's systolic blood pressure (SBP) rapidly decreased to 70 mmHg despite a one-liter bolus of crystalloid and the above interventions.

On admission to ICU the patient's heart rate was 125 bpm, SBP 80 mmHg, temperature 37.8°C, and he was oliguric despite 4 liters fluid resuscitation. A central venous catheter (Cook Medical Inc., IN, USA) was inserted. Central venous pressure was 2 mmHg. The patient's oxygen saturation was 99% on room air, with a respiratory rate of 20 breaths/min. Fluid resuscitation continued with compound sodium lactate and boluses of 4% Albumin. Empiric anti-infective therapy was broadened to piperacillin/tazobactam, ciprofloxacin, flucloxacillin and azithromycin. Intravenous hydrocortisone was administered in stress doses (50 mg i.v. q.i.d.). The patient was drowsy, and continued to complain of neck stiffness. His widespread macular rash persisted.

The patient's only reported past medical history was of atypical migraines. He had not travelled overseas recently. He identified as a man who has sex with men, and denied unprotected intercourse. He had returned a negative HIV test three months previously.

Laboratory tests on ICU admission (Table 1) demonstrated changes consistent with a reactive process with decreased total protein (45 g/L) and serum albumin (22 g/L) levels. Magnesium and phosphate levels were mildly decreased (0.56 mmol/L and 0.26 mmol/L respectively). Serum creatinine and urea levels were normal. Acid-base balance was normal and lactate was mildly elevated (2.2 mmol/L). Repeat cultures and immunoassays were negative. Chest radiography remained unremarkable.

IV noradrenaline was commenced, rapidly rising to 20 mcg/min. By Day 5 in-hospital noradrenaline requirements had risen to 88 microg/min, and vasopressin had commenced at 2 units/hr. CVP remained at 2 mmHg.

12 liters of IV fluid had been administered within 48 hours, but the patient remained oliguric and intravascularly fluid-depleted, confirmed by transthoracic echocardiography that revealed complete collapse of the inferior vena cava and near-oblivation of the ventricular cavities (Online Videos). Creatinine levels rose to 422 μmol/L and urea to 19.9 mmol/L and continuous renal replacement therapy was commenced.

Respiratory virus PCR tests returned negative results

Oxygen saturation remained 100% on ambient air. A vascular access catheter (Cook Medical Inc., IN, USA) was inserted,

The patient complained of right ankle pain on Day 4. Examination revealed mottled limbs bilaterally, and pain on passive movement. Examination of the knees and upper legs were unremarkable.

Vascular ultrasound of the lower legs was normal, with no evidence of a high-resistance waveform. Direct measurement revealed pressures of 60 mmHg and 62 mmHg in the right and left anterior compartments respectively, and 30 mmHg and 31 mmHg in the right and left posterior compartments respectively. Bilateral lower leg anterior and posterior compartment fasciotomies and muscle biopsy were performed.

The patient returned to ICU intubated and ventilated, however worsening arm and upper leg edema were noted. Direct measurement demonstrated pressures >60 mmHg in all compartments.

Table 1 Biochemical parameters, vasopressor requirements and fluid balance during hospital and intensive care admission

In-Hospital Day ^a	Day 0	Day 1	Day 2	Day 3	Day 4	Day 5	Day 6	Day 7	Day 8	Day 9	Day 10	Day 11	Day 12	Day 13	Day 14	Day 15	Day 16	Day 17	Day 18	Day 19
Fluid Input (ml) ^d	9942	3711	2216	14801	5920	2467	9223	3135	4196	3223	2794	2630	2449	1830	433					
Fluid Output (ml)	695	933	1980	7234	5687	9223	6594	6594	7559	5316	3458	2893	2199	981	0					
Daily Fluid Balance (ml)	+9247	+2778	+2336	+7567	+5333	-6756	-3459	-3363	-2093	-2093	-664	-263	+250	+849	+433					
pH ^b	7.36	7.35	7.27	7.24	7.3	7.45	7.46	7.4	7.4	7.41	7.47	7.46	7.46	7.47	7.51					
pCO2 (mmHg)	34	27	39	39	56	36	38	43	38	43	43	36	40	39	35					
Bicarbonate (mmol/L)	25	14	18	17	27	24	26	26	26	23	27	25	27	27	28					
Base Excess	-1	-9	-9	-10	0	0	2	2	1	-1	2	2	3	4	5					
Hemoglobin (g/L)	164	139	144	149	165	59	89	87	89	90	81	74	69	78	71			87	67	73
Hematocrit (L/L)	0.48	0.4	0.42	0.43	0.46	0.18	0.26	0.27	0.25	0.26	0.24	0.22	0.21	0.23	0.21			0.23	0.2	0.21
White Cell Count (x10 ⁹ /L)	22.78	22.43	21.23	32.88	37.91	34.94	12.3	16.87	19.61	16.98	14.09	11.28	11.44	9.86	11.67			10.42	11.32	10.7
Neutrophil Count (x10 ⁹ /L)	21.54	21.88	16.44	32.06	36.95	33.67	11.09	15.66	16.56	9.84	12.24	9.4	8.85	6.94	7.87			6.63	8.74	6.81
Albumin (g/L)	44	34	22	17	14	23	22	24	25	25	24	22	22	22	22			23	24	24
Sodium (mmol/L)	135	138	134	127	130	133	134	136	137	135	136	138	139	138	138			137	137	137
Potassium (mmol/L)	3.7	3.9	3	4.5	5.2	5.6	3.8	4.3	4.2	4.4	4.4	4.2	4	4.4	4.5			5.2	5.2	6
Creatinine (μmol/L)	70	71	121	422	390	350	195	210	193	163	173	184	160	164	184			757	553	765
Urea (mmol/L)	5.8	3.9	7.9	19.9	19.2	18.4	12.5	13.7	13.2	10.6	11.1	9.9	10.7	10.7	9.9			29.2	17.5	23
APTT (secs)	34.9	36.6	40	81.2	78.7	68.2	41.5	43.7	37.5	38.9	41.5	41.5	39.1	39.1	39.2			39.9	37	
Noradrenaline (μg/min) ^c																				
Vasopressin (units/hr)																				

^a In-hospital day; Days highlighted in **bold italics** represent days in ICU.

^b Physiological variables (pH-APTT) represent the most-derived variable in that 24-hour period.

^c Noradrenaline and vasopressor doses are the highest dose required within that 24-hour period.

^d Fluid input and output volumes are for the 24-hour period.

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