Systemic Capillary Leak Syndrome Associated with Compartment Syndrome

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Abstract

Systemic capillary leak syndrome is characterized by recurrent hypovolemic shock attributable to increased systemic capillary leakage. A 26-year-old woman was admitted because of recurrent episodes of hypovolemic shock. Hemoconcentration, hypoalbuminemia, and monoclonal gammopathy were observed. We diagnosed systemic capillary leak syndrome. Three years later, she again had an attack of systemic capillary leak syndrome complicated with pretibial compartment syndrome. This case emphasizes the importance of muscle compartment pressure monitoring during volume resuscitation in patients with systemic capillary leak syndrome.

Key words: hypovolemic shock, hemoconcentration, hypoalbuminemia, monoclonal gammopathy

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Introduction

Systemic capillary leak syndrome (SCLS) is a rare but devastating disorder that was first described by Clarkson et al in 1960 (1). SCLS is characterized by recurrent episodes of increased capillary permeability, resulting in hypovolemic shock due to a marked shift of plasma, up to 70%, from the intravascular to the extravascular space. Laboratory features include hemoconcentration and hypoalbuminemia, often with an associated monoclonal gammopathy without any evidence of multiple myeloma (2). Currently, about 57 cases have been reported, 12 of whom died of the disease during follow-up (3). Development of compartment syndrome by swelling of a massive muscular compartment is an uncommon complication of SCLS (4-6). We describe a case of SCLS associated with pretibial compartment syndrome.

Case Report

In January 2002, a 26-year-old woman was admitted because of shock following a 3-day history of flu-like illness. Three days before admission, she began to have a sore throat. Two days before admission, her body temperature exceeded 38.7°C. On the day of admission, her fever rose to 38.4°C and nausea, general malaise, and lower abdominal pain related to menstruation developed. A month prior, she had had a similar episode. She was admitted because of shock following flu-like illness and menstruation. She recovered with fluid resuscitation and ampicillin/sulbactum and clindamycin administration. On hospital day 18, she was discharged. Toxic shock syndrome or angioedema was suspected. However, the diagnosis was inconclusive.

On physical examination, her temperature was 38.4°C, blood pressure was 84/44 mmHg, and pulse was 120 beats per minute. Her jugular vein was collapsed. Her chest and abdomen were normal. She had not used tampons. On gynecological examination, vaginal infection or pelvic inflammatory disease was not noted.

Laboratory values were as follows: leukocyte count 8.10 × 10⁹/l, hemoglobin 17.5 g/dl; hematocrit 50.8%; platelet count 238 × 10⁹/l; albumin 2.8 g/dl; AST 13 IU/l (normal 12-36 IU/l); ALT 8 IU/l (normal 3-32 IU/l); and creatinine 1.03 mg/dl. Serum electrolytes were normal. Inten-
sive therapy including fluid resuscitation (6.0 l per day), dopamine, ceftazidime and clindamycin administration, and steroid pulse therapy were provided. However, the hypotension did not improve. Generalized subcutaneous edema developed following fluid resuscitation. On hospital day 2, she was still hypotensive and became lethargic. She was intubated. The edema deteriorated dramatically and she developed oliguria. Pulmonary edema was not observed. Serum albumin decreased to 0.9 g/dl. Continuous hemodiﬁlteration was performed. On hospital day 4, urine output increased markedly and edema improved. Blood cultures were negative. Her C1 esterase inhibitor activity was 146% (normal 80-125%). Serum IgG, IgA, and IgM were 1,370 mg/dl, 80.4 mg/dl, and 63.9 mg/dl, respectively. Serum protein immunoelectrophoresis showed paraprotein of the IgG lambda type. SCLS was diagnosed. On hospital day 43, she was discharged. Theophylline (200 mg twice daily) and terbutaline (4 mg three times daily) were administered for prophylaxis against SCLS. However, compliance was poor because of palpitation.

In February 2005, she was again admitted to the hospital because of shock following fever, nausea, and general malaise. Three days before admission, she began to have a fever of 39.6°C. One day before admission, inﬂuenza B was diagnosed. Oseltamivir was administered. On the day of admission, fever rose to 38.0°C and nausea and general malaise developed.

On arrival, her temperature was 38.9°C, blood pressure was 96/46 mmHg, and pulse was 141 beats per minute. She appeared pale and to be in acute distress. Her jugular vein was collapsed. Her chest and abdomen were normal. Edema was not noted.

Laboratory values were as follows: leukocyte count 10.0 × 109/l, hemoglobin 20.7 g/dl; hematocrit 56.9%; creatine kinase (CK) was 187 IU/l (normal 0-180 IU/l); and creatinine 1.04 mg/dl. Liver enzymes (AST and ALT) and serum electrolytes were normal. Fluid resuscitation (6.0 l per day) and dopamine administration were initiated. However, blood pressure decreased to 60/30 mmHg. On hospital day 2, marked generalized subcutaneous edema developed with fluid resuscitation. Hematocrit was 60.7%, albumin was 3.2 g/dl, and CK was 1,111 IU/l. On hospital day 3, albumin decreased to 1.1 g/dl. She was intubated. Over the following days the hypovolemic shock and edema gradually subsided. However, on hospital day 7, pre-tibial compartment syndrome associated with peripheral neuropathy and limitation of dorsal ﬂexion were noted. Measurement of pre-tibial compartment pressure by manometry disclosed 70 mmHg. This pressure measurement can be used to guide treatment, since the risk of ischemic myonecrosis increases markedly as the pressure rises above the mean arterial pressure (4). Fasciotomy should be performed when the tissue pressure rises to within 10-30 mmHg of the diastolic pressure in a patient with any of the other signs or symptoms of a compartmental syndrome (8).

In the present case, acute pulmonary edema was not observed during three SCLS episodes. However, Chihara et al reported the case of SCLS who developed acute pulmonary edema on hospital day 3. They speculate that intravascular overloading accompanied by the recruitment of the initially extravasated ﬂuids and macromolecules resulted in acute pulmonary edema (9). Their case indicates the importance of timely switch from the management of severe hypovolemia to that of acute ﬂuid overload when the recruitment phase starts (10). If kidney function is compromised, hemodialysis or hemodiﬁlteration should be done (10).

We treated the present patient with steroid pulse therapy in the second SCLS episode. Corticosteroid may have a role when cytokine-mediated endothelial damage initiates the capillary leak (9, 10). Tahirkheli and Greipp (2) and Droder et al (11) showed that treatment with terbutaline and theophylline caused episodes of the SCLS to completely abate in some patients and decreased the incidence and severity of episodes in others. Terbutaline and theophylline diminish the increment of capillary permeability induced by bradykinin, by an increase of cyclic adenosine monophosphate (5).

Prolonged survival may provide more time for progression to multiple myeloma. Two patients who progressed to multiple myeloma after the diagnosis of SCLS were reported (12). In patients with monoclonal gammopathy of undetermined signiﬁcance, the actuarial risk of myeloma at 25 years follow-up is 30% and actual risk is 11% (13). Annual surveillance for multiple myeloma is recommended for SCLS patients. In conclusion, this case highlights the importance of muscle compartment pressure monitoring during volume resuscitation in SCLS patients.

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References


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