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Cardiac tamponade as the clinical presentation of Clarkson's disease

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26 y/o man was admitted to the emergency department after repeat episodes of dizziness that resulted in syncopes. Objective examination revealed GCS 15, respiratory rate 16 x/min, pulse 102 x/min, blood pressure 103/70 mmHg, pulse oximetry 98%. Neurological examination did not reveal any pathology.

The first diagnose was unexplained syncope episodes. ECG monitoring and brain CT were performed. It did not reveal pathology. Blood tests revealed hemoconcentration (Er 7,84 milj., Hb 226 g/l, Ht: 67%, Leu27 100). The massive infusion therapy was given i/v, but irrespective of that man remained hypotensive. Repeat blood tests revealed the same hemoconcentration.

The cardiologist was called due to unexplained hypotension. Echo was performed, where the effusion 0,7 – 2,5 cm at the pericardium was found with signs of the cardiac tamponade. The patient was admitted to the cardiac intensive care unit with suspicions of exudative pericarditis unknown etiology. During pericardiocentesis 200 ml of clear serous liquid were acquired. The patients stabilized for some hours, but later the same day patients’ hemodynamics began dropping rapidly down to 70/40 mmHg and increasing doses of noradrenaline and more intravenous fluid had to be introduced.

At the same time oedema started developing on face and extremities, especially legs. The patient developed compartment syndrome with following demyelinating polyneuropathy. The blood sample revealed a low protein level 28 g/l. It was substituted by albumin infusion. The fluid balance was plus 13 410 ml for 34 hours. At that time the preventive continuous renal replacement therapy with ultrafiltration was started. The severe hemodilution with hematocrit drop up to 24.8% was started on the third day after admission.

The diagnosis of Idiopathic systemic capillary leak syndrome (SCLS) or Clarkson’s was made on the fourth day by the exclusion of others causes of systemic capillary leak syndrome as sepsis etc. One of the especially remarkable findings was serum protein analysis positive for paraprotein. On the sixth day, his condition was improved. i/v immunoglobulins were started. The trepan biopsy was performed. It revealed gammopathy. 20 days after admittance patient was discharged in a satisfactory condition.

Idiopathic systemic capillary leak syndrome (SCLS) or Clarkson’s disease is an extremely rare pathology with prevalence less than one in a million. It starts as a sudden increase of capillary wall permeability that results in hypovolemic shock. The fluid that leaves vascular bed is accumulated in third spaces and interstitial tissue and can lead to compartment syndrome, rhabdomyolysis, deep vein thrombosis, and, in rare cases – cardiac tamponade. In "post leak phase" of the disease fluid spontaneously returns to vascular bed causing massive fluid overload with possible cardiopulmonary failure. Cardiac tamponade is a very rare complication of an even rarer disease but should not be forgotten.