Patient with Cold Agglutinin Disease Undergoing Cardiac Surgery Requiring Cardiopulmonary Bypass

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Introduction. Cold agglutinin disease (CAD) is a rare type of autoimmune haemolytic anaemia, caused by cold reactive autoantibodies - cold agglutinins (CA). They react with the red blood cells (RBCs) when the blood temperature drops below normal body temperature. The incidence of CAD has been estimated to be 1/106 per year [Berentsen et al., 2006]. Hypothermic during cardiopulmonary bypass (CPB) and cold cardioplegia (CP) can lead to massive hemagglutination (HA), haemolysis, microvascular occlusion and organ failure.

Aim, Material and Methods. The aim of the case report was to evaluate the possibility of making cardiac surgery in moderate hypothermia conditions for a patient with CAD. In this case report we aimed to describe a patient undergoing aortic valve replacement who was detected to have CAs preoperatively. A brief literature review with the aim of management strategies has been also undertaken.

Results. A 71-year-old man was seen with moderated bioprosthetic aortic valve (Hanckook 23) stenosis. The patient had symptoms related to CAs - anaemia, fatigue, shortness of breath, icterus, although it is hard to interpret symptom associated with CAD because of chronic hepatitis C and chronic heart failure. Although polyspecific warm and cold anti RBC antibodies were detected due IgM drop from 4+ (27.05.) to 2+ (08.07.), we did not administer plasma exchange or other immunologic therapy preoperatively. No information of CAs titter and the exact temperature below which HA due to CA activation occurs was available, cardiac surgery was maintained at 36.2 °C at the lowest nasopharyngeal temperature. In total, 17 ml of warm blood CP was given. The CPB time - 125 min. The highest serum potassium level during CPB time was 6.4 mmol/l. Heartbeat restored spontaneity. Blood products were used in surgery. The patient was weaned successfully from CPB and did require support with inotropes. He received 0.15 µg/kg/min norepinephrine support, which was stopped 90 min after surgery. The patient received 2 RBC concentrates on the first day after the surgery and in total 4 RBC concentrates till the 12th day after the surgery. Troponin I and CK concentration elevation was insignificant. Haemolysis was observed 4 hours after CPB in urine microscopy. First hours after surgery indirect bilirubin concentration were 34 µmol/l, the highest - 57 µmol/l. In urine urobilinogen - 135 µmol/l and bilirubin - 7 µmol/l was detected on the 8th and 12th day after the surgery. No signs of CA-related complications. The patient was discharged at 14 days after the surgery.

Conclusions. Thermal amplitude should be quantified preoperatively. Patients with high titter and high thermal amplitude CAs, need personal planning before cardiac surgery. Core temperature monitoring should be used intraoperatively. Cardiac surgery can be made successfully using normothermic CPB and continuous warm blood CP.