

Hypothermic circulatory arrest for aortic dissection with cryoglobulinemia

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Abstract

Cryoglobulinemia is a cold-reactive autoimmune disease. A 64-year-old man with active cryoglobulinemia presented Stanford type A acute aortic dissection. He had been treated with immunosuppressive drugs and plasma exchange (PE) at our hospital; subsequently, qualitative analysis of cryoglobulin (CG) was negative. He underwent emergency ascending aorta replacement using cardiopulmonary bypass (CPB) under deep hypothermia circulatory arrest with selective cerebral perfusion. The total CPB time, aortic cross clamp time, and selective cerebral perfusion time were 255, 153, 56 minutes, respectively, and the minimal nasopharyngeal temperature was 17.3°C. Our patient had no significant perioperative complications. Hence, if PE is performed appropriately and CG is negative, patients with cryoglobulinemia who exhibit severe preoperative symptoms can safely undergo surgery with deep hypothermia.

Introduction

Cryoglobulinemia is a clinical disorder characterized by the presence of cryoglobulins (CGs) in the serum. CGs are immunoglobulins (IGs) that precipitate in vitro at temperatures below the normal body temperature (<37°C) and redissolve upon warming. Cardiovascular surgeries, such as cardiopulmonary bypass (CPB) usually requires systemic hypothermia, has a risk of organ disorders due to microangiopathy induced a rapid increase in blood viscosity. We present a case of ascending aorta replacement with CPB under deep hypothermic circulatory arrest in a patient who experienced acute aortic dissection during cryoglobulinemia treatment.

Case report

A 64-year-old man was referred to our hospital 6 months ago for cryoglobulinemia. He had previously been treated for gangrene of the right toe 2 years prior during winter; however, since then symptoms of necrosis and numbness of the lower extremities worsened. CGs were detected through qualitative analysis (Fig. 1a). However, no additional abnormalities were detected upon further examination. Hence, he was diagnosed with active idiopathic cryoglobulinemia. Subsequently, immunosuppressive therapy with corticosteroids and cyclophosphamide was initiated. However, he had persistent numbness in the lower limbs, and CG levels slightly decreased (Fig. 1b) without a negative outcome. Therefore, he underwent three rounds of plasma

exchange (PE) and double filtration plasmapheresis; subsequently, qualitative analysis of CG was found to be negative (Fig. 1c). The day after PE, our patient experienced sudden chest and back pain. Contrast-enhanced computed tomography revealed Stanford type A acute aortic dissection.

Regarding the use of hypothermia during surgery, since the patient had undergone PE the day before, we decided to perform the surgery using CPB under deep hypothermia circulatory arrest. After esophageal temperature was lowered to 20°C and circulatory arrest was accomplished, selective cerebral perfusion was initiated. Cardiac arrest was induced using retrograde blood cardioplegia at 20°C, which was administered every 30 minutes. We finished anastomosing the distal aorta, and then, started recirculation and rewarming. After anastomosis of the proximal aorta, antegrade blood cardioplegia was administered at 30°C to 35°C. Weaning from CPB was smooth and uneventful, with no changes in cardiac function. Also, no signs of agglutinin reaction and thrombosis in the CPB circuit were observed. The total CPB time, aortic cross clamp time, and selective cerebral perfusion time were 255, 153, and 56 minutes, respectively. The minimal nasopharyngeal temperature during the procedure was 17.3°C.

One day after surgery, the patient was extubated in the intensive care unit and had no significant postoperative complications. Six months after surgery, he received outpatient treatment for cryoglobulinemia.

Discussion

CGs are proteins that become insoluble and typically precipitate at temperatures below normal body temperature and dissolve back into the serum upon rewarming CGs can be either IGs or a mixture of IGs and complement components¹. In open heart surgery, systemic hypothermia, and cold or tepid cardioplegia procedure using CPB are typically used to protect the myocardium and internal organs. However, in patients with cryoglobulinemia, these methods may have deleterious effects. Regrettably, there are no guidelines for the management of patients with CG during CPB. Therefore, we searched an extensive literature and found only 13 cases of CPB in patients with cryoglobulinemia.

The profiles and treatment of the 14 cases, including our case, are presented in Table 1 and 2. The severity of preoperative CG was based on various conditions. Among these patients, especially, our patient had very severe symptoms and needed intensive treatment for cryoglobulinemia (Table 1).

In many cases, surgery was performed using systemic normothermia (>35°C) (Table 2). However, since Stanford type A dissection may require total arch replacement, we had to lower the body temperature of our patient. The literature review revealed that only 3 cases have been reported which used moderate hypothermia (25~27°C), two of them underwent PE previously for decreasing the CG levels before surgery^{2,3}.

In cardioplegia, warm cardioplegia via antegrade was typically used, and two of 3 cases used moderate hypothermia were not used blood cardioplegia^{2,3}. In our case, myocardial protection was performed with intermittent retrograde cold blood cardioplegia during hypothermia. During these procedures, there may be a risk of thrombosis of the coronary veins. Hence, after proximal anastomosis, the temperature was increased approximate 35°C and antegrade warm cardioplegia was initiated for flushing thrombus.

Fortunately, in our case, the qualitative analysis result was negative for CG after PE therapy. Hence, surgery could be performed under deep hypothermia and cold cardioplegia, even for the patient with severe cryoglobulinemia symptoms.

Conclusions

We successfully performed aortic replacement under hypothermic circulation arrest in a patient with acute aortic dissection during cryoglobulinemia treatment and obtained favorable results. Our patient underwent PE a day before CPB and was negative for CG. Thus, if PE is performed appropriately and CG is negative, then CPB under hypothermia can be considered in patients with cryoglobulinemia who exhibit severe preoperative symptoms.

Conflicts of interest . The authors declare that they have no competing interests.

Authors' contributions . All authors contributed to the writing of the final manuscript.

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Figure legends

Qualitative analysis of cryoglobulin (CG). (a) Upon admission to our hospital and (b) during immunosuppressive treatment with corticosteroids and cyclophosphamide, CG levels slightly decreased without a negative outcome. (c) After three rounds of plasma exchange (PE) and double filtration plasmapheresis. CG was found to be negative.

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