Successful treatment of a pregnant woman with Marfan's syndrome with aortic prosthetic valve thrombosis

Chao A, Chao AS, Wu I-Hui
National Taiwan University Hospital, Taipei, Taiwan

Objective

Valve obstruction is a serious complication associated with potential lethal complication. Treatment of prosthetic valve thrombosis includes administration of a fibrinolytic agent or surgery. Here, we present the successful resuscitation of a pregnant patient with Marfan's syndrome, who had undergone Bentall's procedure who developed aortic prosthetic valve thrombosis and cardiogenic shock in her third trimester.

Methods

Retrospective case report.

Results

A 37 year-old pregnant patient experienced several episodes of near syncope in the morning at 29 weeks gestation. The patient was diagnosed with Marfan's syndrome at the age of 29 with Type I dissecting aortic aneurysm and severe aortic regurgitation when she suffered from severe chest pain radiating to her back. . She underwent Bentall's operation with a St Jude mechanical valve. Three years later, another dissection occurred in the mid-thoracic aorta and she underwent thoracic aortic grafting. Anticoagulant was shifted from oral warfarin to subcutaneous low molecular weight heparin, enoxaparin since the first trimester. The patient was sent to our emergency department (ED). She suddenly developed pulseless electrical activity in ED admission. Cardiopulmonary resuscitation was started and emergency cesarean section was done at ED. Despite returned of spontaneous circulation, her hemodynamics was unstable under high doses of inotropes. Veno-arterial extracorporeal oxygenation was initiated. Because of massive bleeding from vagina and wound, she was rushed to operating theater for hysterectomy. During the operation, transesophageal echocardiography showed poor motility of prosthetic aortic valves and thrombi in left atrium and ventricle. Urokinase was administered, with 4, 400 U/kg/hour intravenously, which resulted in severe wound and vagina bleeding. Massive blood transfusion and re-laparotomy were performed for hemostasis. Hemodynamic improvement was noted and inotropic agents were tapered within one day. A bedside echocardiography showed complete lysis of the thrombus. Urokinase was stopped after 20 hours use. Prior to removal of the ECMO cannulae, fluoroscopy was performed which showed bilateral symmetric mobility of aortic leaflets. The total time for circulatory support by ECMO was 23 hours. The patient was left with a left sided hemiplegia. Brain CT showed multiple wedge-shaped infarcts at bilateral fronto-parietal lobes, left occipital lobe and cerebellum. Seven days after ECMO removal, extubation was done and four days later she was transferred to general ward with oral warfarin. Her baby did not survive. After rehabilitation, the patient can walk without any aid and she is regularly followed up in cardiovascular clinic.

Conclusion

Pregnancy in a Marfan's syndrome patient poses high risks to both mother and fetus. This is a rare and complicated case who presented challenges faced by the patient herself who has been treated with Bentall's operation and thoracic aortic grafting and her physicians as well. During pregnancy the selection of anticoagulant and its dosage are paramount. The balance between protecting the prosthetic valve from thrombosis in a hypercoagulable state and the prevention of major bleeding in a patient with the potential of aortic dissection is difficult. Perhaps more meticulous care needed; more frequent echocardiography for assessing the aorta and thrombus formation in the prosthetic valve should be done. Just as important is the circulatory support from ECMO, making the use of urokinase possible to the moribund patient and circumventing the high risk open heart surgery.