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Transcatheter Aortic Valve Implantation in Bicuspid Aortic Valve Patients with Coagulation Factor 7 and 11 Deficiency and Atrial Fibrillation

CASE REPORT

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INTRODUCTION

Transcatheter aortic valve implantation (TAVI) is now a standard therapy for symptomatic severe aortic stenosis (AS) patients who have high or intermediate surgical risk.[1] Positive data for low-risk patients are also plentiful for the tricuspid valve population. On the other hand, the anatomical challenges specific to bicuspid aortic valve (BAV) disease do not make TAVI easy or understandable in this subset.[2] However, the long-term observations after TAVI in BAV patients are not yet available. Patients with BAV may require aortic valve replacement at an earlier age than those patients with tricuspid aortic valve (TAV). Considering that TAVI indications expand to younger and healthier patients, long-term observation of TAVI in BAV is of great importance. As a less invasive approach compared to surgical aortic valve replacement, TAVI has less major hemorrhagic complication rates.[3] Co-existing bleeding disorders can further complicate cardiovascular surgical and interventional procedures. Although acquired von-Willebrand factor deficiency is one of the most common bleeding disorders in TAVI patients, physicians may encounter severe AS patients with different coagulation factor deficiencies as the procedure is progressively being applied in a larger patient population.[4]

Factor 11 plays a role in intrinsic coagulation cascade and there are several mutations responsible for its deficiency (hemophilia C) which is a very rare condition with an incidence of 1 in a million in general population. Severe factor 11 deficiency is defined as <20% factor 11 activity.[4] In addition to that congenital factor 7 deficiency is also a very rare autosomal recessive bleeding disorder with an incidence of 1:500 000. Clinically significant hemorrhagic complication rates tend to increase if the factor 7 activity is below 8%-10%.[5]

There have been numerous reports on literature for the management of bleeding disorders like hemophilia A-B or von Willebrand factor deficiency during cardiovascular surgical procedures.[5] In our knowledge, this is the first case in the literature regarding a patient with BAV stenoses with combined factor 7 and 11 deficiency undergoing a successful TAVI procedure.

CASE REPORT

A 53-year-old male patient has been admitted to our clinic with complaints of palpitation, syncope, and subsequent chest pain. Patient's electrocardiogram (ECG) at the emergency department showed narrow QRS tachycardia with 180 beats per minute. After implementation of intravenous diltiazem control, ECG showed atrial fibrillation (AF) with 112 beats per minute. Bedside transthoracic echocardiography (TTE) showed AS with bicuspid aorta and mean gradient of 36 mm Hg. Left ventricular systolic functions were normal. After that, coronary angiography was performed, and severe stenotic lesions were found on left anterior descending artery, obtuse margin branch, and first diagonal branch. Following coronary angiography, amiodarone and beta blocker therapy was initiated and the patient

Table 1. Patient's Coagulation Test Levels

Laboratory Test	Patient's Results	Normal Values
Coagulation Factor 7 activity	0.9%	60%-150%
Coagulation Factor 11 activity	2.4%	60%-150%
International normalized ratio	2.3	0.9-1.3
activated partial thromboplastin time (aPTT)	93.5 seconds	21-32 seconds
Coagulation Factor 5 activity	119%	50%-150%
Coagulation Factor 2 inhibitor	87%	50%-150%
Coagulation Factor 10 activity	102%	50%-150%
Coagulation Factor 9 activity	128%	50%-150%

returned to sinus rhythm. After maintaining sinus rhythm, detailed TTE revealed severe AS with a mean gradient of 42 mm Hg and type 0 BAV.

The patient was evaluated by the heart team, and preoperative preparation was initiated with anticipation of coronary artery bypass grafting and surgical aortic valve replacement. Perioperative routine test results showed international normalized ratio (INR) of 2.3. The patient was not using any anticoagulation medication and there was no abnormality in liver functions. A detailed hematological evaluation revealed that the patient had congenital deficiency of coagulation factors 7 and 11. Blood results regarding coagulation factors are summarized in Table 1. The patient had no history of uncontrollable bleeding and had not undergone any major surgical procedure up to that time.

After the detection of Factor 7 and 11 deficiency, the patient was re-evaluated by heart team collaborating with a hematologist, and surgery was deemed high risk especially for bleeding. As a less invasive approach, the patient was scheduled for TAVI procedure. Informed consent of patient was obtained before the procedure. Computed tomography was performed for TAVI measurements (Figure 1). Replacement of the deficient factors is planned before the procedure. As there are not any Factor 11 concentrate available in our country, we planned to replace the patient with 10 mL/kg of fresh frozen plasma (FFP). Also, recombinant factor 7a (Novoseven, Novo Nordisk, Denmark) was provided and held in the operating room to be given in case of emergency. Because of the short half-life of coagulation factors, FFP replacement was done just before the procedure.

Transcatheter aortic valve implantation procedure was carried out toward the left common femoral artery. Femoral artery was cannulated by roadmap technique and 2 proglides were premounted. Baseline activated clotting time (ACT) level was 220 seconds, and 2000 units of heparin was given in order to hold the ACT level at approximately around 300 seconds. The procedure was done under mild sedation without any complication. Edwards SAPIEN 3 29 mm valve is implanted (Supplementary Video 1), and the patient was discharged after 2 days of uneventful follow-up.

DISCUSSION

Although there are numerous case reports in literature regarding patients with coagulation factor deficiencies undergoing cardiac operations, this is the first case defining a BAV stenoses patient with combined factor 7 and 11 deficiencies undergoing successful TAVI procedure.[7] Replacement of the factors with either FFP or specific

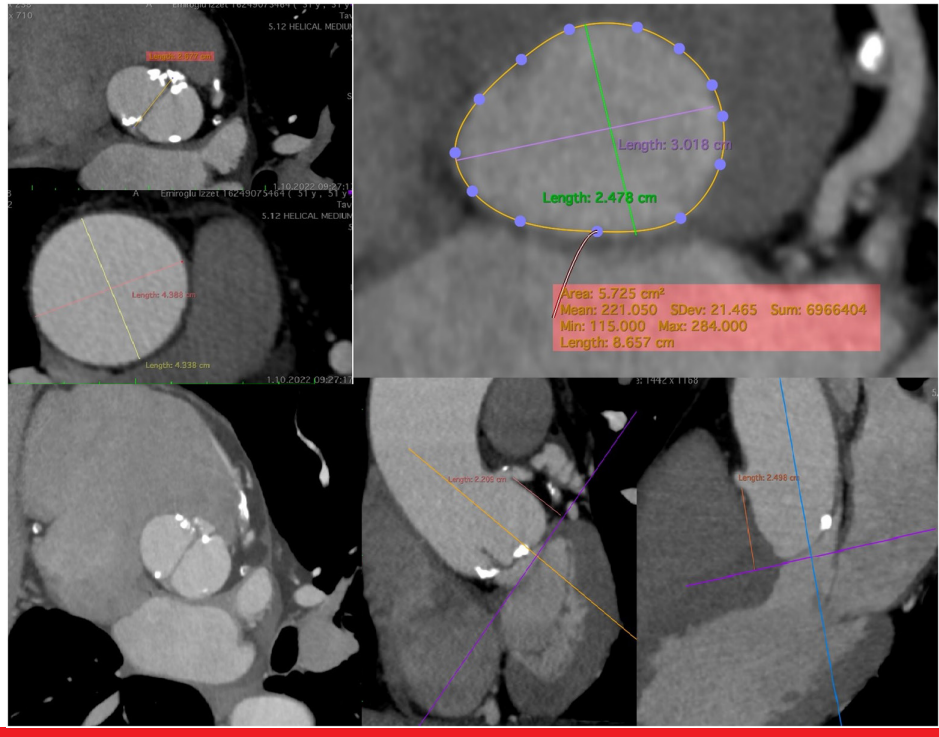

Figure 1. Computed tomography image of the bicuspid aortic valve measurements.

coagulation factor concentrates is the preferred strategy before surgical procedures as reported in the previous case reports.[5] In the light of our hematology consultants' suggestions, we performed the procedure after replacing the patient with FFP (10 mL/kg). Regarding the short half-life of coagulation factors, we applied FFP just before the procedure. Another option is to replace the patient with deficient factors only, but as Factor 11 concentrate was absent in our country, we only provided Factor 7 concentrate ready in case of emergency. A meticulous planning of the procedure was done, and according to CT images, we decided to perform TAVI operation from the left common femoral artery as there was a fibrous plaque on right common femoral artery. Cannulation of the left femoral artery was obtained with roadmap images.

The patient had obstructive coronary artery disease, but the initial complaints were attributed to severe AS by the heart team so the aortic valve replacement procedure was prioritized and symptom-guided intervention strategy is adopted for the coronary artery disease in order to avoid unnecessary interventions and usage of antithrombotic agents, which will increase the bleeding risk. Regarding the possibility of future coronary interventions, we decided to implant a balloon-expandable aortic valve, and considering the young age of the patient, we selected Edwards SAPIEN 3 valve which has satisfactory data about long-term durability.[8]

Another problem about the patient was decision of whether to start anticoagulation medicine or not as the patient was first admitted with AF. As having paroxysmal AF, coronary artery disease, and bioprosthetic heart valve, the patient needs anticoagulation therapy in normal circumstances. Our hematology consultants decided that the patient does not require additional anticoagulation medicine as the basal INR values are in the range of 2-3. Although there is a lack of sufficient data regarding these group of patients, there are reports that endogenous thrombin potential (ETP) values of patients with severe hemophilia A can resemble the levels of patients on oral anticoagulation.[9] Endogeneous thrombin potential levels can be used as a surrogate marker for coagulable state and thrombotic risk.[10] There are opinions in literature to manage hemophilia patients who need anticoagulation according to the levels of the deficient factors.[11] Considering the low levels of factors 7 and 11 in our patient, the decision not to start oral anticoagulation is compatible with the literature. Our plan is to follow the INR levels closely and start low-dose warfarin if it decreases below 2.0. Catheter ablation of AF is an option in case of recurrent AF episodes.

In this case report, we presented a successful TAVI procedure in a patient with severe AS, BAV, paroxysmal AF, and factor 7-11 deficiency and medical management strategies at the follow-up.

Informed Consent: Informed consent was obtained from the patient.

Supplementary Video 1: Coronary angiography, the implantation of the Sapien 3 valve and aortography.

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