An 86-year-old woman with polycythemia vera (high allele burden of JAK2 V617F mutation: 95.3%) was referred for pulmonary edema due to severe aortic valve stenosis and was scheduled for transapical transcatheter aortic valve replacement. Because hydroxyurea administration was interrupted because of retromalleolar ulcers, preoperative blood examinations showed severe thrombocytosis (1,500 × 10^9/l) and neutrophilic leukocytosis (24.6 × 10^9/l). Multiplate impedance aggregometry showed a slight increase in platelet aggregation with the ASPItest and the ADPtest and aggregation within normal limits with the TRAPtest (all by Roche Diagnostics, Rotkreuz, Switzerland); thromboelastometry showed increased maximal clot firmness with the FIBTEM test and normal MCF with the EXTEM and with the INTEM tests (TEM International GmbH, Basel, Switzerland). The patient was not receiving anticoagulant agents or antiplatelet drugs, because an extremely high platelet count may lead to hemorrhagic complications because of acquired von Willebrand disease (1). Transapical transcatheter aortic valve replacement with a SAPIEN 3 valve bioprosthesis (Edwards Lifesciences, Irvine, California) was performed with a good final result. Six hours later, the patient’s hemodynamic status suddenly deteriorated, necessitating cardiopulmonary resuscitation. Transesophageal echocardiography showed massive thrombosis of the aortic valve bioprosthesis with severely reduced leaflet excursion (Figure 1A, Online Video 1). Percutaneous femorofemoral extracorporeal membrane oxygenation was positioned, but in a few minutes, acute thrombosis of the oxygenator occurred, necessitating complete heparinization and emergency replacement of the whole circuit. Coronary angiography was performed and showed patent coronary ostia with distal coronary embolization in the left anterior descending coronary artery (Figure 1B, Online Video 2). Despite maximal support, the patient died a few hours later of massive cerebral hemorrhage. At autopsy, the leaflets of the SAPIEN 3 bioprosthesis were blocked by massive white thrombosis, on both the aortic and ventricular sides (2–4) (Figure 1C). The bioprosthesis was well seated in the aortic root, without either coronary ostia...
obstruction or paravalvular leaks. On histological examination, the thrombus consisted predominantly of platelet aggregates in a fibrin network (Figure 1D). Occluding platelet emboli were found in the distal intramural small vessels of the anteroseptal ventricular myocardium.

REFERENCES


KEY WORDS polycythemia vera, transcatheter aortic valve replacement, valve thrombosis

APPENDIX For supplemental videos, please see the online version of this article.