A serious complication of high-power short-duration radiofrequency atrial fibrillation ablation associated with genetic hypodysfibrinogenemia

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Introduction

Cardiac tamponade is a rare and potentially fatal outcome in patients who undergo atrial fibrillation (AF) ablation.[1] In the United States about 2.7 million are known to have AF; and of those, approximately 0.8% undergo catheter ablation. Cardiac perforation with radiofrequency ablation (RFA) likely occurs through 2 mechanisms: mechanical complications with the sheath and catheter or steam pops with RFA. Surgery to repair the cardiac perforation is more common after a steam pop.[2] High-power short-duration (HPSD) RFA is now commonly used in AF ablation owing to shortened procedure times with no decreased long-term efficacy or increased complications compared to low-power long-duration ablation.[3–6] This case highlights the occurrence of cardiac tamponade during AF ablation using HPSD RFA in a patient with a rare bleeding diathesis owing to hypodysfibrinogenemia. This appears to be the first known case report of a patient with the Kagoshima mutation and hemopericardium during AF ablation. This case highlights whether fibrinogen levels and repletion should be assessed in a patient with persistent bleeding and whether screening for rare bleeding disorders in Asian patients prior to ablation may help assess potential risks.

Case report

A 61-year-old male patient of Japanese origin with hypertension, peripheral vascular disease, and symptomatic paroxysmal AF despite antiarrhythmic drugs elected for catheter ablation. The patient was not on anticoagulation prior to the procedure and denied prior surgeries, trauma, or history of bleeding. During HPSD radiofrequency ablation (50 W for 10 seconds) with the TactiCath™ (Abbott Catheters, Abbott Park, IL) Contact Force Ablation Catheter, Sensor Enabled™ of the left atrial appendage (LAA) ridge near the junction of the anterior aspect of the left superior and inferior pulmonary veins, the patient had an impedance rise and steam pop. Hypotension was noted and intracardiac echocardiography confirmed a large pericardial effusion. Despite heparin reversal with protamine and emergent pericardiocentesis, there was continued rapid reaccumulation of pericardial blood. Blood product replacement with 6 units of packed red blood cells, 4 units of fresh frozen plasma, and 4 packs of platelets was administered. With no cessation of bleeding, the patient was transferred to the operating room for emergent sternotomy. Persistent bleeding from a 5-mm-size perforation at the base of the LAA was found and repair of this area with LAA ligation was performed. He was placed on low-dose heparin after the surgery and later transitioned to oral anticoagulation. The patient recovered well from surgery and was discharged 3 days later. However, despite 15 units of

KEY TEACHING POINTS

- An underlying bleeding disorder may exacerbate cardiac complications such as pericardial effusion and tamponade during atrial fibrillation ablation.
- Assessing a bleeding panel and checking fibrinogen levels may be reasonable in cases of persistent bleeding.
- Genetic testing is reasonable in susceptible populations with a bleeding diathesis of unclear etiology.

KEYWORDS Hypodysfibrinogenemia; Atrial fibrillation; Radiofrequency ablation; Tamponade; Kagoshima mutation
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cryoprecipitate repletion, persistently critically low fibrinogen levels at less than 60 mg/dL multiple days and weeks after surgery were noted. Genetic testing was remarkable for a mutation on the *FGG* gene (FGG c.1019C>T, p.(Thr340Ile). This mutation, termed the Kagoshima mutation, is a known cause of hypodysfibrinogenemia.

Discussion

Hereditary fibrinogen abnormalities are divided into 2 subclasses: firstly, quantitative fibrinogen deficiencies, or type I, in which there are low (hypofibrinogenemia) or absent (afibrinogenemia) plasma fibrinogen antigen levels; and secondly, qualitative fibrinogen deficiencies, or type II, in which there are normal (dysfibrinogenemia) or low (hypodysfibrinogenemia) antigen levels with low functional activity.[7] Hereditary dysfibrinogenemia is characterized by the synthesis of abnormal fibrinogen that leads to reduced functional capacity.[7] It is mostly asymptomatic but can also present as bleeding, thrombosis, or both.[8] Hypodysfibrinogenemia is the least reported of congenital fibrinogen disorders with unknown prevalence, as patients are generally asymptomatic, although mild-to-moderate bleeding and some cases with thrombosis due to impaired fibrinolysis have been reported.[7,8] Interestingly, in our patient, further work-up demonstrated that he had the Kagoshima genetic mutation, where there was an amino acid substitution from threonine-314 to isoleucine that resulted in impaired fibrinogen function and hypofibrinogenemia. This missense alteration is rare and is reported in several unrelated individuals in Japan. Patients with this mutation have defective binding of thrombin to abnormal fibrin that leads to increased free thrombin levels caused by abnormalities in the low-affinity thrombin-binding site or to resistance to fibrinolysis. This causes very low clot stability, delayed cross-linking of patient fibrin-catalyzed calcium ions, and delayed cross-linking of patient fibrin catalyzed by activated factor XIII.[9]

Our patient did not have a personal or family history of bleeding disorders or venous thromboembolism prior to AF ablation. The rapid reaccumulation of pericardial blood during the procedure leading to hemodynamic instability was concerning for a bleeding diathesis and prompted further work-up confirming the Kagoshima mutation as his cause of hypodysfibrinogenemia. This mutation was not associated with consumption, as the fibrinogen levels would go up to 100 with cryoprecipitate. Possible explanations for our case are that the initial injury of cardiac perforation during ablation did not clot owing to hypodysfibrinogenemia or the cardiac perforation was so large, or was in a location such that surgery was inevitable. Our HPSD ablation protocol is fairly standard, and the literature does not appear to suggest increased steam pops with this approach.[10] To potentially minimize steam pops, ablation can automatically be stopped if the programmed temperature and impedance parameters are breached. Although fresh frozen plasma does contain fibrinogen, the amount per volume is approximately 10 times less than that of cryoprecipitate.[11] Given his critically low baseline fibrinogen level, cryoprecipitate would have been the optimal choice and might have assisted with clotting and mitigated the need for surgery. However, the thrombosis risk with cryoprecipitate administration acutely following AF ablation vs the surgical risk should be considered for each patient profile.

Conclusion

This case highlights that an underlying bleeding disorder may exacerbate the known complication of cardiac perforation associated with HPSD RFA. In circumstances where persistent bleeding occurs it may be reasonable to assess a bleeding panel and assess for fibrinogen levels.

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