Primary antiphospholipid syndrome (PAPS) is a disease manifested by a tendency toward recurrent arterial and venous thrombosis, placental thrombosis, placental thrombosis leading to recurrent fetal loss, thrombocytopenia, and a prolonged activated partial thromboplastin time (PTT) in the absence of other autoimmune diseases\(^1\)\(^3\)\(^7\)\(^8\)\(^9\) in these patients. Because of the hypercoagulable state in this disease, long-term anticoagulation therapy is recommended, although the choice and optimal dose of anticoagulant is not known.\(^1\)\(^4\)

Inferior vena caval filter placement has been recommended to prevent recurrent pulmonary emboli in high-risk patients\(^5\)\(^6\) (ie, free-floating or poorly adherent proximal thrombus on venogram, chronic pulmonary hypertension, or marginal respiratory reserve). To our knowledge, the use of an inferior vena cava filter to prevent recurrent pulmonary embolism in patients with PAPS has not been reported. Moreover, periprocedural management strategies to prevent postoperative thromboembolism have not been defined. We report a difficult and ultimately fatal case of PAPS in a patient with a previously placed vena cava filter after uncomplicated cholecystectomy despite treatment with heparin and aspirin.

**CASE REPORT**

A 61-year-old Hispanic woman was diagnosed as having PAPS when she presented with a history of chronic leg ulcer, recurrent spontaneous abortions, pulmonary emboli, extremely high titers of anticardiolipin IgG and IgM, and no other autoimmune diseases.\(^7\) In November 1990, she was admitted to St. Joseph Hospital at the Creighton University Medical Center with recurrent pulmonary emboli that were confirmed by pulmonary angiography. A bird’s nest filter (Cook Inc, Bloomington, Ind.) was placed in the inferior vena cava to prevent further pulmonary emboli. Her clinical condition temporarily stabilized and the platelet count normalized after treatment with low-dose heparin, 5,000 U subcutaneously twice daily, and aspirin, 100 mg/d (Fig 1), and she was discharged from the hospital on this regimen.

She was readmitted to the hospital 7 months later with acute cholecystitis and she underwent cholecystectomy that evening. Following cholecystectomy, she developed progressive thrombocytopenia and required 2 U of packed red blood cells for blood loss.
Low-dose heparin treatment was restarted immediately postoperatively. Aspirin treatment was withheld for 2 days then restarted again on the third hospital day (Fig 1). On the fifth postoperative day, she had been walking around the nurse’s station when she developed acute shortness of breath, diaphoresis, tachycardia, and peripheral cyanosis. An emergency pulmonary angiogram showed large segmental perfusion defects in the lingula and posterior aspects of the upper lobe of the left lung and a large occlusive thrombus in the descending branch of the right pulmonary artery. Cather-directed thrombolysis was attempted with 250,000 IU of urokinase, diluted 50,000/10 ml of sterile water, injected over a period of 10 min. After a 10-min interval, an additional 500,000 IU were administered in a similar manner. Repeated contrast imaging showed only minimal improvement in perfusion of the lung. Repeated blood gas measurements showed no change in oxygenation. She became hypotensive and bradycardic during the procedure and died en route to surgery for an emergency embolectomy. At autopsy, both recent and organized thromboemboli within recanalizing pulmonary vasculature were noted. One possible source of thrombus included the inferior vena caval bird’s nest filter, which showed both recent and organized thrombus formation extending proximally from the filter (Fig 2).

**DISCUSSION**

Insertion of an inferior vena caval filter has been recommended to prevent recurrent pulmonary emboli. The most frequent indication for filter insertion is a contraindication to anticoagulation, followed by failure of anticoagulation to prevent embolism. Some patients with PAPS have extremely prolonged activated PTT and prothrombin time (PT) as in the case presented. This patient had marked elevations in PT (21.6; normal range, 10.9 to 13.1 s) and PTT (136.4; normal range, 24.8 to 37.4 s) before anticoagulation therapy, which made monitoring with the clotting time tests on therapy impossible. At the time of her second episode of pulmonary embolism, we empirically treated this patient with low-dose heparin (5,000 U subcutaneously twice daily) and aspirin (100 mg/d) as recommended by Sammaritano et al11 and Alarcon-Segovia and Sanchez-Guerrero.7 In addition, we placed a vena caval filter in an attempt to prevent recurrent pulmonary emboli. Unfortunately, pulmonary embolism recurred despite this regimen. We speculate that the hypercoagulable state with its high tendency toward local thromboembolism formation makes the inferior vena caval filter less useful in preventing pulmonary embolism in this group of patients.

Although treatment with perioperative corticosteroids, anticoagulants and/or antiplatelet agents appears to protect against postoperative thrombotic complications in patients with lupus anticoagulants,10,11 its necessity in nonvascular procedures such as cholecystectomy is controversial. We did not treat this patient with corticosteroids because coagulation parameters did not change after a short-term course of prednisone treatment during her previous hospital admission (Fig 1). Moreover, the therapeutic role of steroids in patients with PAPS is questionable.12 We doubt that the addition of steroids in our patient would have changed the clinical course.

In summary, prophylactic treatment with low-dose heparin and aspirin in this patient with PAPS with a previously placed vena cava filter failed to prevent postoperative thrombotic complications. We believe that consideration should be given to more aggressive anticoagulation therapy to avoid postoperative thromboembolism in these patients, particularly in those patients with a previously placed vena caval filter.

**REFERENCES**

5. Greenfield IJ, Michna BA. Twelve-year clinical experience with the Greenfield vena cava filter. Surgery 1988; 104:706-12