Phlegmasia Cerulea Dolens: Rare Complication of Vena Cava Filter Placement in Man With Paraplegia

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Abstract

Objective: To describe a complication of placement of an inferior vena cava (IVC) filter in a man with paraplegia.

Design: Case report.

Participants/Methods: A 48-year-old man with T11 paraplegia secondary to an L1 burst fracture underwent thoracic spinal fusion. The postoperative course was complicated by deep vein thrombosis (DVT) of the right common femoral vein, which was treated with warfarin.

Results: During rehabilitation, the hematocrit declined, and fluctuance was noted along the surgical site. Computed tomographic scan suggested a hematoma in the paraspinal and latissimus dorsi muscles. Warfarin was discontinued, and an IVC filter was placed. He subsequently developed severe leg pain, followed by hypotension, acute renal failure, and compartment syndrome in bilateral lower extremities requiring fasciotomies. Ultrasound and computed tomographic angiogram showed extensive bilateral lower extremity DVTs and pulmonary emboli. The diagnosis of cerulea dolens was made. Mechanical and pharmacological thrombectomy was aborted secondary to bleeding complications and hypotension. The patient died shortly after care was withdrawn at the family's request. The autopsy revealed multiple thrombi in IVC, bilateral pelvic and femoral veins, and left pulmonary artery embolus, consistent with phlegmasia cerulea dolens.

Conclusions: Inferior vena cava filters may prevent pulmonary embolism but do not affect the underlying thrombotic process. An IVC filter should be recognized as a possible thrombogenic nidus in patients with spinal cord injury who have known DVT.

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Key Words: Phlegmasia cerulea dolens; Venous thrombosis; Spinal cord injuries; Vena cava filters; Pulmonary embolism; Paraplegia

INTRODUCTION

Phlegmasia cerulea dolens (PCD) is a rare but potentially fatal complication of iliofemoral thrombosis. In this condition, extensive deep vein thrombosis (DVT) causes complete occlusion of venous outflow from a large portion of the leg, leading to increased interstitial tissue pressure, decreased capillary blood flow, and tissue ischemia (1). It has been reported after traumatic hip dislocation, in the presence of associated conditions (eg, neoplasm, cardiovascular disease, infection), with the use of oral contraception, and as a complication of an inferior

vena cava (IVC) filter (1–3). We are unaware of reported cases of PCD either in individuals with spinal cord injury (SCI) or in individuals with Günther Tulip IVC filters. Phlegmasia cerulea dolens presents with tense swelling of a limb, cool extremities, and absent distal pulses. Treatment options are limited, and if they are not instituted expediently, this condition can lead to gangrene of the extremities or, ultimately, death (1). We present a case of severe PCD in an individual with paraplegia.

CASE PRESENTATION

A previously healthy 48-year-old African American man sustained a L1 burst fracture that resulted in a T11 ASIA A SCI. The patient received the standard methylprednisolone protocol. He underwent T12–L2 posterior spinal fusion with L1 corpectomy with anterior plating via anterolateral approach at the 11th rib on postinjury day

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Figure 1. Computed tomography of abdomen showing hematomas (arrows) in the paraspinal muscles and adjacent to the left latissimus dorsi muscle.

(PID) 1. The perioperative course was complicated by acute tubular necrosis, hyponatremia, prostate injury due to indwelling catheter, and *Staphylococcus* bacteremia from presumed intravenous line contamination. Heparin for DVT prophylaxis was started on PID 2. Thrombocytopenia (platelet count of $69 \times 10^3/\mu\text{L}$) of unclear etiology was noted on the same day. On PID 5, the patient developed right common femoral vein DVT while still receiving heparin. Heparin-induced thrombocytopenia was ruled out. His past medical history and family history were negative for coagulopathy. The patient was started on enoxaparin for the treatment of DVT on PID 5. Warfarin was started on PID 9, and enoxaparin was to be discontinued once therapeutic levels of warfarin were achieved.

The patient was transferred to the acute SCI inpatient rehabilitation unit on PID 12. He was participating well in the rehabilitation program, when his hematocrit dropped from 27.0 to 22.5% and he developed associated fatigue and tachycardia. On physical examination, there was notable fluctuance along the surgical site. Computed tomographic scan of the abdomen and pelvis revealed a hematoma at T12 level (Figure 1). At that point, warfarin and enoxaparin were discontinued, and a Günther Tulip IVC filter (Cook Medical, Bloomington, IN) was placed by an interventional radiologist on PID 14. The patient continued to participate in the rehabilitation program, and his course was unremarkable except for bilateral lower extremity dysesthetic pain, which was treated with gabapentin with subjective improvement.

On PID 23, his leg pain worsened considerably. His blood pressure decreased to 83/54 mmHg, and his heart rate increased to 110 beats/min. He had no complaints of chest pain or shortness of breath, and his pulse oxygen saturation was stable at 97 to 98% on room air. Despite an intravenous normal saline fluid bolus of 500 mL, his systolic blood pressure continued to drop to into the 50–mmHg range. His physical examination was significant for increased leg circumference bilaterally. The patient remained alert and responsive, and his chest examination remained clear. His laboratory workup showed an increase of serum creatinine from 0.9 to 3.4 mg/dL, an increase of blood urea nitrogen from 29 to 45 mg/dL, an increase of potassium from 4.6 to 6.7 mmol/L, and a decrease in bicarbonate from 21 to 11 mmol/L within 24 hours. His arterial blood gas showed a pH of 7.41, pO_2 of 142 mmHg, pCO_2 of 14 mmHg, and base excess of 8.7 on 5 L of oxygen. A code was called, and the patient was intubated for airway protection. He was transferred to the medical intensive care unit and started on neosynephrine and dopamine vasopressors. All blood cultures were negative. Renal artery thrombosis was considered, but the renal sonogram was negative.

His blood pressure continued to decrease despite treatment with multiple vasopressor agents. Of most concern, the patient developed tense distention of his lower body below the groin level, and his feet were cool and poorly perfused. Bilateral lower extremity compartment syndrome was diagnosed, and the patient underwent 4 compartment fasciotomies on PID 25. On the same day, abdominal ultrasound showed arterial and venous renal flow in the right kidney and only venous flow in the left kidney. Left renal artery flow was not well visualized but was seen in the computed tomographic angiography performed at a later time. Ultrasound of the lower extremities showed DVTs in the bilateral common and superficial femoral veins and the bilateral popliteal veins. A venogram showed extensive DVTs in bilateral common femoral veins. A hypercoagulable workup (anticardiolipin antibodies, lupus anticoagulant, Factor V Leiden, Prothrombin G20210A, heparin-induced thrombocytopenia antibody, and serotonin–release assay) was negative.

On PID 27, computed tomography of the abdomen and angiogram showed:

1. No definite evidence for retroperitoneal or paraspinal hematoma.
2. Absence of contrast from the IVC, iliac veins, and common femoral veins consistent with acute thrombosis to the level of the IVC filter (Figure 2).
3. Filling defects within the pulmonary arteries consistent with bilateral pulmonary embolism (Figure 3).
4. Moderate left and small right pleural effusion.
5. Extensive diffuse anasarca.

The patient continued to have progressive multiorgan failure with renal failure (creatinine peaked at 6.5

Figure 2. Computed tomography angiogram showing the Günther Tulip filter (arrow) and no distal blood flow in the inferior vena cava (arrow).

Figure 3. Computed tomography angiogram (coronal view) showing a clot in the left pulmonary artery (arrow).

mg/dL), hypotension despite maximal doses of 5 vasopressors, and respiratory failure while on mechanical ventilation. Hemodialysis was begun on PID 26. On PID 28, as a last resort, interventional radiology service attempted tissue-plasminogen-activator infusion and mechanical thrombectomy (Figure 4), but it was discontinued prematurely due to bleeding from the fasciotomy sites. His white blood cell peaked at $42.9 \times 10^3/\mu\text{L}$ on PID 30. The patient's family decided to withdraw care on PID 31, and the patient passed away shortly after. The autopsy showed the following:

1. S/P L1 burst fracture with T11 paraplegia.
2. S/P IVC filter placement.
3. Thrombosis of IVC and bilateral pelvic and femoral veins.
4. Left lower lobe pulmonary embolus.

Figure 4. Venogram of common femoral vein obtained during attempted thrombectomy showing extensive deep vein thrombosis.

5. Pulmonary edema.
6. Bilateral pleural effusions.
7. Evidence of autolysis in the abdominal organs.

DISCUSSION

Venous thromboembolism is a very common complication of SCI (4–6). It is a cause of pulmonary embolism after SCI, which can ultimately cause death. The second edition of Clinical Practice Guidelines for thromboembolism prevention published in 1999 noted a 40% rate of DVT in patients with acute SCI (6). A review article by Green noted the incidence of DVT to be 48 to 100% without anticoagulation prophylaxis and approximately 10% with prophylaxis (4). The incidence of pulmonary embolism has been reported to be 2.6%, and it is the cause of death in 10% of individuals with SCI in the first year postinjury and in 3% after the first year (4).

Venous thromboembolism in SCI is thought to be not only caused by venostasis but also due to a transient hypercoagulable state, reduced fibrinolytic activity, increased blood factor VIII activity, dehydration, and/or concomitant injury to the soft tissue or long bone (6). In addition to SCI, other clinical risk factors for DVT are lower extremity fracture, previous thrombosis, cancer, heart failure, obesity, and age greater than 70 years, none of which this patient had prior to his SCI (6). Another recent study by Jones and colleagues reported other risk factors for venous thromboembolism to include male gender, complete paraplegia compared with

complete tetraplegia, and being African American (7). Other significant comorbidities include metastatic cancer, underlying chronic neurological disease, history of psychiatric disease, and depression/anxiety. In this case, the significant risk factors were ethnicity, gender, and complete paraplegia.

There is a potential for DVT to progress proximally and to embolize in a high percentage of the cases (6). However, routine use of prophylactic IVC filter does not seem to be indicated as a preventive measure against pulmonary embolism after SCI (5,8). Those patients with high risk factors, such as long bone fractures, DVT despite prophylactic anticoagulation, and contraindications to anticoagulation, may be appropriate candidates for prophylactic IVC filters (4,8). In this case, the patient had the last 2 indications for IVC filter placement, that is, he developed his initial DVT despite being on anticoagulation; then he developed hematoma at the surgical site and had a contraindication for anticoagulation at that time.

The term “phlegmasia cerulea dolens” (La phlebite bleue) was first used by Gregorie in 1938 when he described a condition that included venous thrombosis, painful ischemia, and purple discoloration (1). Phlegmasia cerulea dolens is caused by complete occlusion of venous outflow from an extremity secondary to acute massive thrombosis, and it should include occlusion of both the major outflow channels and many of the collateral veins. This venous occlusion causes abnormally elevated intravascular hydrostatic pressure, which in turn causes massive extravasations of fluid into the surrounding tissue (9). This extravasation leads to a decrease in arterial pressure. Combined with excessive interstitial hydrostatic pressure, the arterial closing pressure is exceeded, which collapses the arterial wall. These events ultimately cause tissue ischemia (9). There is a subsequent loss of vascular volume, which can exceed 3 to 5 L and can result in hypotension, which was seen in this case. Signs and symptoms of PCD include intense pain, edema, decreased pulses, and a cyanotic, mottled extremity. It can be associated with a compartment syndrome, as seen in this case. Mortality from PCD has been reported in 20 to 40% of cases, and amputation has been performed in 12 to 50% of survivors (1,10–12).

Goals of the treatment are to restore normal volume, prevent venous gangrene, and prevent pulmonary embolism (9). Treatment of PCD remains controversial given its rarity. Immediate measures that could be taken are rapid volume replacement, extreme elevation of the extremity, and fasciotomies if compartment syndrome is present (9). Other treatment options include sympathetic blockade, sympathectomy, venous surgical thrombectomy, catheter-directed thrombolysis, anticoagulation, and amputation (1). In this case, anticoagulation and catheter-directed thrombolysis were attempted without success. Although there is very little literature on the subject of PCD, thrombectomy has been thought to be the most effective treatment option when it is initiated

early (1,13). In 2 reported cases of PCD, a filter was placed via internal jugular vein to prevent pulmonary embolism from a subsequent thrombectomy (3,9).

This case illustrates a rare but potentially devastating complication of PCD with IVC filter placement. It is felt that there are 2 possible mechanisms for the development of PCD with IVC filter placement: thrombi propagate distally from the filter after the trapping of an embolus or proximally from the site of insertion of the filter in the femoral vein to the filter. Inferior vena cava filters have many other potential associated complications. These include bleeding or formation of thrombus at the site of insertion; perforation of the vena cava; and malposition or migration of the filter, including migration of the filter to the heart (1,2,4,14). One of the common long-term complications of IVC filters is caval thrombotic occlusion, which is reported to occur in 6 to 30% of cases (5,15,16). Postphlebitic syndrome from chronic venous occlusion also has been reported with IVC (5). Percutaneous IVC filter placement may prevent pulmonary embolism, but it does not affect the underlying thrombotic process and may contribute to thrombosis, as seen in this case (5,17).

In this patient, a retrievable Günther Tulip filter was placed. This has been used as a retrievable filter in Europe since 1992, and it has been approved by the United States Food and Drug Administration as a permanent filter since October 2000 and as a retrievable filter since October 2003 (16,18). It is a nonmagnetic conical-shaped filter with 4 main struts and 8 additional filter wires and a hook at the apex that is used in the retrieval process. This filter is 45-mm long and has an expanded diameter of 30 mm. It can be placed via the femoral or jugular approach, but it can only be retrieved from the jugular approach (16). Like any other filter, the Günther Tulip filter causes a mild fibrotic reaction that eventually fixes the device to the endothelium. Allen reported only 1 complication in 53 cases, which was the inability to retrieve the filter (18). Others reported complication rates of 3.6% for recurrent pulmonary embolism and 6% for total caval occlusion (16). We are unaware of any report of PCD as a complication of a Günther Tulip IVC filter.

CONCLUSIONS

This 48-year-old man died as a result PCD as a secondary complication of traumatic spinal fracture and paraplegia. Physicians who use IVC filters should be aware of the risk of PCD and be alert for possible signs and symptoms so that prompt diagnosis can be made and the appropriate treatment initiated early.

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