

Case in Point

Pulmonary Vein Thrombosis Associated With Metastatic Carcinoma

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A patient with severe onset abdominal pain was diagnosed with pulmonary vein thrombosis in the setting of concurrent cancer diagnoses.

Pulmonary vein thrombosis (PVT) is rare and underdiagnosed in clinical practice.

It has been described following lobectomy, lung transplant, and in association with metastatic carcinoma.¹⁻⁴ Some cases have been described as idiopathic.⁵⁻⁷ Its exact incidence is unknown, and treatment depends on etiology. On the other hand, pulmonary artery thromboembolism is a well-known entity with identified risk factors as well as clearly defined modalities of management. The following is a case of PVT, which occurred in the setting of small cell carcinoma of the lung (SCLC) and mantle cell lymphoma of the small intestine.

CASE PRESENTATION

A 66-year-old male veteran with a past medical history of type 2 diabetes mellitus, hypertension, and chronic obstructive pulmonary disease, who had a 40 pack-year history of cigarette smoking, was admitted to the hospital for severe, sudden onset abdominal pain. The pain was

localized in the right lower quadrant and then became generalized. It was sharp, aggravated by movement, and relieved by rest. The patient reported being constipated for the past couple of days.

A review of systems revealed that he had been coughing for about 3 days prior to admission. A computed tomography (CT) scan of the abdomen showed pneumoperitoneum and a mass with mural thickening around the distal ileum/cecal area (Figure 1). There was also a partially visualized mass in the infrahilar area of the right lower lobe and bilateral adrenal masses seen on the scan. A chest CT with contrast was then performed, which showed a 7.5 x 6.6 x 6.6-cm mass in the right lower lobe posterior to the right hilum. The mass encased the right mainstem bronchus, and there was a low-density-filling defect in the inferior branch of the right pulmonary vein (Figure 2). An echocardiography did not show any thrombus within the atria or ventricles.

The patient underwent emergent exploratory laparotomy for bowel perforation. The operative finding was a small perforation of the small intestine with an associ-

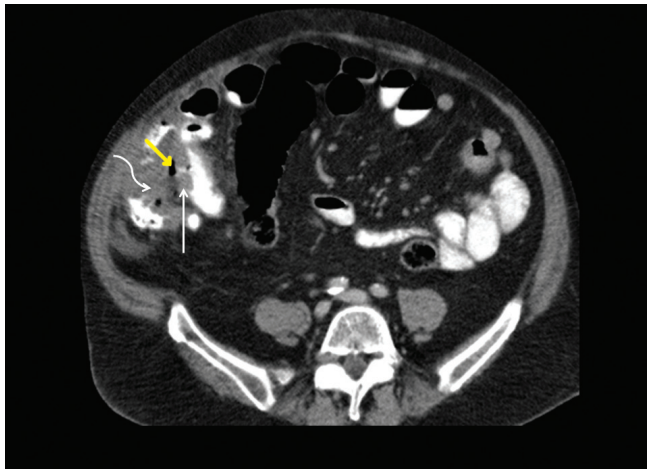
ated mass. There were metastatic lesions throughout the abdomen. A partial small bowel resection was performed. Post exploratory laparotomy, a fiberoptic bronchoscopy was performed, which revealed a 1-cm fungating lesion at the takeoff of the superior segment of the right lower lobe. Brushings were obtained from the mass. The pathology of the lung mass was small cell carcinoma, whereas that of the bowel mass was mantle cell lymphoma. Brain magnetic resonance imaging revealed that he had metastasis to the brain with a 4-cm mass in the cerebellum. He was anticoagulated with heparin for the PVT. Based on his poor functional status and his overall clinical condition, his prognosis was poor. He received hospice care and died 3 months later.

DISCUSSION

Pulmonary vein thrombosis is a rare condition. The incidence is unclear, as most of the literature includes case reports. The majority of PVT cases are reported following lobectomy for malignancy and lung transplantation.¹⁻³ The incidence following lung transplant was reported in the early postoperative period to be 15% in a center during the first 2 years of the

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Figure 1. Computed Tomography of the Abdomen—Coronal



Mural thickening in the distal ileum (straight arrow) and adjacent cecum (curved arrow). Free air (yellow arrow).

Figure 2. Computed Tomography of the Chest With IV Contrast—Axial



Filling defect in the right inferior pulmonary vein (arrow).

study.³ Pulmonary vein thrombosis has also been described following metastatic cancer, such as liposarcoma.⁴

This patient's case was discovered in the setting of SCLC and mantle cell lymphoma of the small intestine. Small cell carcinoma of the lung was reported to invade the pulmonary vein into the left atrium.⁸ In this patient, the left atrium was not invaded. There have been cases of spontaneous or idiopathic PVT described, presenting as abdominal pain, hemoptysis, and chest pain.⁵⁻⁷ No precipitating causes were detected in these patients.

The pathogenesis of PVT from a tumor is unclear, although several theories have been postulated: It could result from direct extension of the tumor into the vein, from compression of the vein by the tumor, or from epithelial damage as a result of tumor invasion. The tumor thrombus has been described to extend into the right atrium.^{6,8} The mechanism of thrombosis remains unclear in the patient postlobec-

tomy or postlung transplantation, although intraoperative torsion and injury of vessels are implicated. Similar to deep vein thrombosis, PVT could also result from intimal damage or sluggish flow in the pulmonary stump in the postoperative patient.^{2,9,10}

The presentation of PVT is usually nonspecific, including dyspnea, cough, pleuritic chest pain, and hemoptysis. It has been reported as causing massive hemoptysis due to acute pulmonary infarction.⁷ Acute PVT occurring postoperatively in the lung transplant patient may be disastrous and lead to early postoperative allograft failure.¹¹ Pulmonary vein thrombosis may also present more insidiously with recurrent pulmonary edema and pulmonary fibrosis.¹² This patient presented with abdominal pain; further workup led to the finding of a lung mass. Pulmonary vein thrombosis has been reported to result in systemic emboli, resulting in cerebrovascular accidents, or it can manifest as aseptic and tumor emboli.^{2,5,10,13,14}

Newer CT techniques have made identifying PVT possible in a similar manner to which pulmonary arterial emboli are detected by using the pulmonary venous phase of a contrast CT of the chest.⁵ Echocardiography may demonstrate the extension of the thrombus into the atrium; a transesophageal echocardiogram would be preferable over a transthoracic echocardiogram. Magnetic resonance imaging of the chest is another useful modality for diagnosis, because it is able to distinguish between a bland thrombus and a tumor thrombus in the pulmonary vein.¹⁵

Treatment of PVT depends on the overall clinical condition of the patient. Irrespective of the etiology, a review of the literature does not indicate the preferred duration of anticoagulation or preference for modality of anticoagulation between oral vitamin K antagonists or heparin—low molecular or unfractionated.^{1,3-6} Patients who develop PVT following malignancy are usually anticoagulated with therapy for the cancer. The treatment of PVT

in the setting of lung transplant is more challenging and includes systemic heparinization, thrombolytics, and surgical thrombectomy.^{3,11,16} The majority of the literature includes case reports with varying morbidity and mortality, depending on the etiology. Ninety-day mortality of 38% was reported following lung transplant.³

CONCLUSION

Pulmonary vein thrombosis presents in a nonspecific manner. The diagnosis is now more readily made with the advent of a variety of diagnostic modalities, especially with transesophageal echocardiography, which may be performed at the bedside in the intensive care unit. The treatment remains challenging with mortality dependent on the etiology. A diagnosis of PVT needs to be considered in patients with appropriate risk factors. A high index of suspicion will enable the diagnosis in the proper clinical scenario. ●

Author disclosures

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