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CASE REPORT

Idiopathic pulmonary vein thrombosis: An unexpected cause of respiratory distress and acute heart failure. A case report and review of the literature

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ABSTRACT

Introduction: Pulmonary vein thrombosis is a potentially fatal disease. The association between pulmonary vein thrombosis and chronic heart failure has not been described in the literature.
Case report: Herein, we report a case involving a 73-year-old woman, with a medical history of ischemic congestive heart failure, who was hospitalised for acute decompensated heart failure with respiratory distress. A computed tomography pulmonary angiography was performed to rule out the possibility of pulmonary embolism, and it showed evidence of pulmonary vein thrombosis. No cause was determined for the pulmonary vein thrombosis; hence, it was considered idiopathic and anticoagulation therapy was initiated for the patient. However, the patient died a few days after admission to the intensive care unit.
Conclusion: This case of pulmonary vein thrombosis is presented to promote awareness of this disease entity. We also want to emphasize the importance of maintaining a high index of clinical suspicion for this diagnosis, particularly in patients with acute decompensated heart failure who are refractory to standard therapy.

African relevance

- Pulmonary vein thrombosis is a potentially lethal condition.
- The presentation of pulmonary vein thrombosis is usually non-specific.
- Pulmonary vein thrombosis can lead to both cardiac failure and respiratory distress.
- A high index of clinical suspicion is needed for appropriate diagnosis.

Introduction

Pulmonary vein thrombosis is a potentially lethal condition that is often underdiagnosed or undetected. There have been several reports of pulmonary vein thrombosis; however, none of these reports have delineated a relationship between pulmonary vein thrombosis and chronic heart failure. Specific factors that precipitate a heart failure hospitalisation can often be identified, although studies suggest that 40%–50% of acute decompensated heart failure episodes have no known cause [1]. When precipitants are identified, it is imperative that they are treated appropriately, and that effective interventions are developed to improve the management of decompensated heart failure. We present a case of PVT that occurred synchronously with known chronic heart failure, and resulted in decompensated heart failure with acute respiratory decline.

The present case was observed in a district hospital located in the south east of Tunisia. Available resources include several university departments. The radiology department, with easy access to the different structures, has a Computed Tomography (CT) scan unit (32-slice multidetector CT). The majority of laboratory tests are available, with the exception of procalcitonin and lactate assay, among others.

Case report

A 73-year-old woman presented to the emergency centre with a seven-day history of progressive dyspnea, orthopnea, and pleuritic chest pain. Her medical history included type 2 diabetes and ischemic congestive heart failure. On physical examination, she

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appeared pale and was in moderate respiratory distress, as indicated by the use of her accessory muscles of respiration. Her vital signs included an oxygen saturation of 77% on room air, blood pressure of 110/65 mmHg, heart rate of 122 beats per minute, respiratory rate of 30 breaths per minute, and temperature of 37.9 °C. Auscultation of her lungs revealed coarse crackles to the mid-zones bilaterally. Her electrocardiogram showed normal sinus rhythm. The initial arterial blood gas analysis performed on room air revealed a pH of 7.44, a pCO_2 of 35 mmHg, and a pO_2 of 46 mmHg. The laboratory analysis revealed anaemia with a haemoglobin level of 9.8 g/dL (normal: 11. 5–15), leukocytosis of $14.7 \times 10^3/mm^3$ (normal: 4. 0–10 $\times$ 10^3), an elevated C-reactive protein (CRP) level of 75 mg/dL (normal: 0–6), and an N-Terminal pro-B-type natriuretic peptide (NT pro-BNP) level of 11.000 pg/mL (normal value <400). Her cardiac-enzyme levels were found to be normal. Serum electrolytes, coagulation, hepatic and renal analysis were within normal range. No growth was observed at microbiology investigations of sputum. Three samples from blood cultures were also negative. The chest radiograph showed interstitial thickening and cardiomegaly. The patient was treated for a presumed infective decompensation of her chronic heart failure and she received diuretics, intravenous antibiotics (amoxycillin-clavulanic acid with levofloxacin), and continuous positive airway pressure (CPAP) therapy. Over the next several hours, the patient's clinical status continued to deteriorate; hence, she was transferred to the intensive care unit (ICU) and further investigations were undertaken. Transthoracic echocardiography revealed left ventricular hypokinesis and systolic dysfunction (ejection fraction of 40%), moderately elevated pulmonary pressure, normal left atrium size, and no intracardiac mass. Additional blood tests revealed an elevation in the fibrin D-dimer level. The rate of cardiac enzymes has not changed. Arterial blood gas analysis (under 100% oxygen through a non-rebreather mask) showed a pH of 7.39, a pCO2 of 37 mmHg, and a pO_2 of 59 mmHg. Initially, we approached this case as an acute pulmonary embolism, but the computed tomography pulmonary angiography (CTPA) revealed a large thrombus obstructing the left superior pulmonary vein, bilateral pleural effusions, and right pulmonary condensation (Fig. 1[A and B]). This unusual incidental finding prompted further investigation into potential underlying predisposing factors, including a thrombophilia screen, antinuclear antibodies, and tumour markers (alpha-fetoprotein, beta 2 microglobulin, carcino-embryonic antigen, CA 19-9), all of which were within normal limits. An extended CT scan of the abdomen and pelvis was negative for malignancy. Based on these findings, the pulmonary vein thrombosis was thought to be idiopathic, and anticoagulation therapy with low-molecular weight heparin was started on the fifth day of the hospitalisation. Antibiotic therapy, initiated at admission, was continued. However, the patient's condition deteriorated despite further treatment with invasive mechanical ventilation and inotropic therapy. The patient died of multi-organ failure approximately ten days after admission to the intensive care unit.

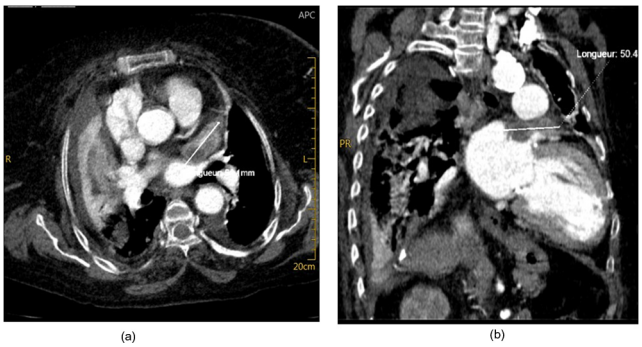

Fig. 1. Computed tomography pulmonary angiography. (Top) Axial image showing a 50-mm thrombus obstructing the left superior pulmonary vein with bilateral pleural effusion and a right pulmonary condensation. (Bottom) Sagittal image showing a large thrombus in the left superior pulmonary vein extending into the left atrium.

Discussion

Pulmonary vein thrombosis is a rare but potentially serious condition. In adults, acquired pulmonary vein obstruction can have various causes, including radiofrequency catheter ablation, pulmonary neoplasm, surgical complications of lung transplantation or lobectomy, and fibrosing mediastinitis [2–4].

In the present case, the etiology of the pulmonary vein thrombosis was not identified; hence, it was considered idiopathic. Few reports have described idiopathic or "spontaneous" pulmonary vein thrombosis. However, several cases have been reported since 2012, due to the development of powerful imaging techniques such as CT and high-resolution CT. According to a retrospective study conducted on 57 Japanese patients with chest pain (age = 73.8 ± 8.6 years), using 64-slice multi-detector CT scans, pulmonary vein thrombi were clearly demonstrated in 35 patients (61%). H. Takeuchi concluded that pulmonary vein thrombosis was not rare in elderly patients with chest pain and no clear predisposing factors, and that pulmonary vein thrombosis might be more common in older individuals [5].

The presentation of pulmonary vein thrombosis is usually nonspecific, making the clinical diagnosis more challenging. Depending on the acuity of the pulmonary venous obstruction and the presence of venous collaterals, pulmonary vein thrombosis can present acutely with cough, dyspnoea, pleuritic chest pain, and haemoptysis, or as a progressive deterioration in respiratory function. As a result, it is common for it to be misdiagnosed as pulmonary embolism. To our knowledge, few cases of idiopathic PULMONARY VEIN THROMBOSIS presenting with respiratory distress have been described in the literature. In contrast with patients in earlier studies, in which the occurrence and severity of the symptoms were confined to patients with severe and multiple stenosis [6], our patient developed severe symptoms even though she only had one occluded pulmonary vein.

Additional examinations such as chest radiograph, electrocardiogram (ECG), and arterial blood gas (ABG) laboratory analysis are nonspecific and may be misleading. Common radiographic findings include multifocal opacities, nodular lesions, pleural effusions, and interstitial septal thickening [7]. Chuang-Chi Liaw et al. prospectively studied pulmonary vein thrombosis in 222 cancer patients. Radiological pulmonary abnormalities were detected in more than 80% of the cases. D-dimer and crp levels were elevated in 89% and 95%, respectively [8].

There is no gold standard for diagnosis of pulmonary vein thrombosis, and usually a combination of diagnostic modalities is required. Indeed, the best diagnostic test is chosen according to the context. Echocardiography may be advantageous in critically ill patients since it is performed at the bedside. Transesophageal echocardiography would be preferable over a transthoracic echocardiography due to the relative close proximity of the pulmonary veins to the distal oesophagus, but it may be more invasive, sometimes requiring sedation. CT scan is often the initial means of discovering pulmonary vein thrombosis, as the mode of presentation usually mimics pulmonary embolism. The 64-slice multidetector CT seems to be reliable for the detection of pulmonary vein thrombosis. Furthermore, a longer scan delay after contrast administration, by using the pulmonary venous phase, may reduce flow artefacts and allow better evaluation of the pulmonary veins [9].

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