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Chronic Pulmonary Embolism Mimicking Pulmonary Angiosarcoma

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We present the case of a 41-year-old woman who presented with chest pain. CT for possible pulmonary embolism showed a mass filling the left main pulmonary artery, with imaging features suggestive of pulmonary angiosarcoma. Open thoracotomy for excision of the mass revelaed no mass, only evidence of a chronic pulmonary thrombus of the left pulmonary artery. We conclude that the radiologic features of chronic pulmonary thrombus may mimick those of pulmonary angiosarcoma.

Case Report

A 41-year-old woman presented to the emergency department complaining of a three-day history of left-sided chest pain. The pain was described as pressure-like, pleuritic, made worse with ambulation and when supine. It was constant, increasing in intensity and not associated with any alleviating factors, hence her request for evaluation. She had denied any history of trauma, fevers, cough, night sweats nor hemoptisis. She denied any dyspepsia, and noted no relief of her symptoms with over-the-counter antacids.

Six weeks prior to the onset of her chest pain she had complained of right hip pain, radiating down the leg. This was diagnosed as radicular pain secondary to sciatica for which she had been under bed rest and analgesics. This slowly improved, and she had reinitiated full ambulation about two weeks preceding her presentation.

Her past medical history was remarkable for depres-

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Abbreviations: CT, computed tomography; FDG, fluorodeoxyglucose; PET, positron emission tomography

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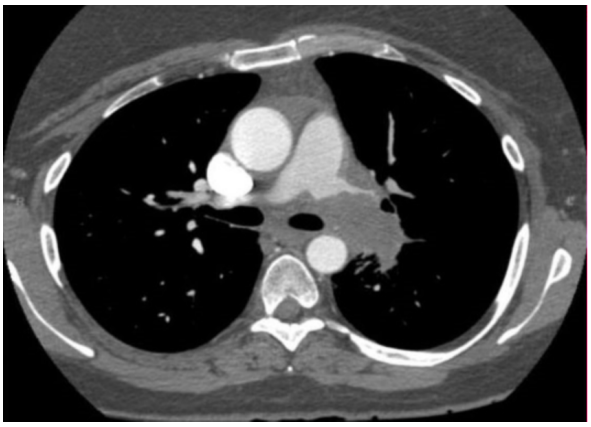

Figure 1A. 41-year-old woman with chronic pulmonary embolism. Axial CT image of the chest demonstrating a mass extending outside the vessel wall with a large filling defect involving the left main pulmonary artery and extending into all sub-branches.

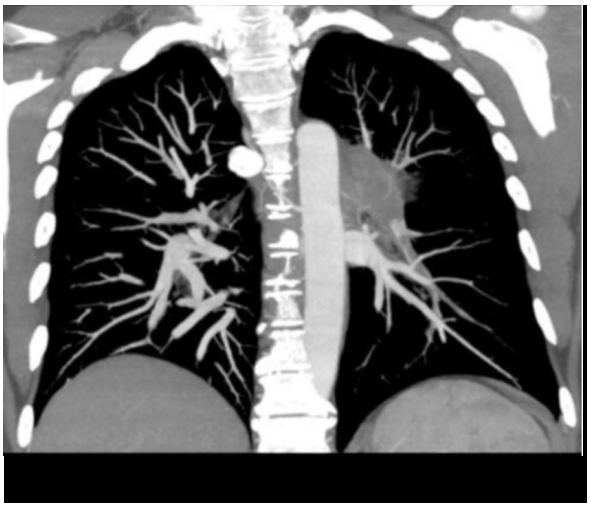

Figure 1B. 41-year-old woman with chronic pulmonary embolism. Coronal reformatted CT image of the chest demonstrating a mass extending outside the vessel wall.

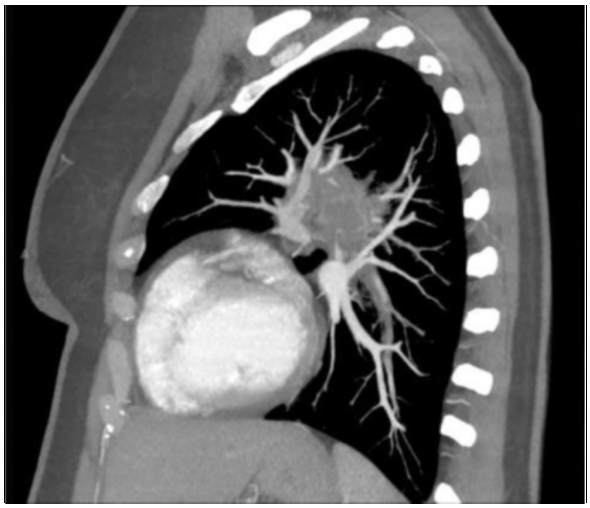

Figure 1C. 41-year-old woman with chronic pulmonary embolism. Sagittal reformatted CT image of the chest demonstrating a mass extending outside the vessel wall.

sion and occasional migraine headaches, both of which had been controlled. She was gravida 2, status-post two cesarean sections, the last of which was performed five years prior. She was started on oral contraception five months earlier, Yaz®, in addition to Topiramate and Escitalopram. She denied any life-long toxic habits. Her family history was remarkable for lymphoma in her mother. She had no known family history of coagulopathy or other hematological conditions.

Upon presentation to the hospital she was evaluated for her chest pain by way of cardiac enzymes and complete blood count, which were both normal. An initial chest radiograph was also noted to be normal, as was an EKG. Nonetheless, a CT scan of the chest using a protocol to evaluate for a pulmonary embolism was performed which was remarkable for a large filling defect involving the left main pulmonary artery and extending into all sub-branches, most pronounced in the left lower lobe (Fig. 1). There appeared to be a mass extending outside the vessel wall. In addition, there was evidence of focal infiltration into the adjacent lung parenchyma with pre and post contrast Hounsfield units of 15 and 60, respectively. This was suggestive of a pulmonary angiosarcoma.

In view of the pulmonary artery occlusion, she was admitted and closely monitored. She had an unremarkable hospital course, which included a negative work up for coagulapathy. With improvement of her chest pain, she was sent home on anticoagulation with warfarin. An FDG-PET scan was later performed, which did not reveal any FDG-avid lesions. A subsequent bronchial-alveolar lavage and pulmonary function testing were also unrevealing. Interestingly, she underwent open thoracotomy for excision of the left thoracic mass, and at the time of surgery no mass was found. There was only evidence of a chronic pulmonary embolism of the left pulmonary artery which was evacuated. Following her surgery, she developed pleural effusions managed with diuretics. Due to progressive dyspnea, even at rest, she was reevaluated in the emergency department. A repeat CT scan of the chest was then positive for bilateral pleural effusions, greater on the left, with adjacent atelectasis, but no evidence of pulmonary embolism. A thoracentesis was performed with cytology negative for malignancy and findings consistent with a transudate.

Discussion

Pulmonary embolisms present acutely, usually as a complication of a deep leg thrombosis, renal, or pelvic thrombus. Larger thrombi may cause hemodynamic compromise by their location at the bifurcation of the main pulmonary artery or the lobar branches. Smaller thrombi may travel distally and are often associated with

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pleuritic chest pain, presumably by initiating an inflammatory response contiguous to the parietal pleura. Most pulmonary emboli are multiple, with the lower lobes being involved in the majority of cases.

Variations may occur within the fibrinolytic qualities of the pulmonary vasculature which may result in chronic, persistence of pulmonary embolisms [1]. Undoubtedly the inciting event is an acute thrombotic episode, but the exact mechanism or susceptibility of patients to have persistent thromboembolisms is unknown. Small-vessel arteriopathy is believed to be the culprit, later in the course of the disease, which may contribute to the progression of hemodynamic compromise [2]. This may eventually lead to clinical manifestations, such as progressive dyspnea and pulmonary hypertension.

The incidence of chronic pulmonary embolism has been estimated as 0.1-3.8% of patients with a prior, venous thromboembolism [3-4]. but many of these patients have an unidentified acute event. The theory regarding possible chronic inflammation, predisposing the vessel to develop persistent thrombus has previously been proposed. This may be possible through a coagulopathic state which may impede resolution of a clot, as is the observation in myeloproliferative states or splenectomized patients [4].

The most common presentation is dyspnea, followed by pleuritic chest pain. It is therefore quite common to find patients being evaluated, initially by chest radiographs with non specific findings. These may range from normal to irregular vascularity or pleural thickening. In addition, some patients are submitted to pulmonary function testing, which is usually normal. However, up to 20% may present with a restrictive pattern, which may lead to misdiagnosis of interstitial lung disease.5 Nonetheless, the differential diagnosis is likely to lead to further imaging techniques.

The diagnostic approach with imaging for thromobembolic events has consisted of CT pulmonary angiography. However, despite its relative availability, the diagnosis may be missed due to flow artifacts. Enhancement of chronic PE may make the diagnosis difficult to distinguish from that of pulmonary angiosarcomas [6], as both may enhance with contrast. In fact, clinical correlation is usually indistinguishable for either condition. However, one may be vigilant for images of lobulated lesions or extravascular extension with angiosarcomas as opposed to chronic PE on CT scans [7]. As previously described, even chest radiographic findings such as hilar/pulmonary artery enlargement, pulmonary nodules and pleural effusions may favor a diagnosis of angiosarcomas of the lung [7].

In differentiating chronic pulmonary embolisms from angiosarcomas, several methods have been used. These include contrast enhanced CT, magnetic resonance imaging (MRI) and FDG-PET scans. The latter may be noteworthy as angiosarcomas are usually FDG-avid, while organized thrombus typically do not show increased uptake [8]. Ultimately, needle and endovascular biopsies are the proposed modalities to make a final diagnosis of pulmonary angiosarcoma [9].

In this particular case, the differential diagnosis was that of a chronic pulmonary embolism versus an angiosarcoma. In fact, her findings were so suggestive of a mass, that she underwent thoracotomy for resection. Surprisingly, she had a chronic embolism which was removed, but this was as unexpected occurrence due to its location; it was unilateral.

It is important to discuss that unilateral central pulmonary arterial embolism is rare [10], but when found should be differentiated from pulmonary artery sarcoma. In this case description, the initial finding of a large filling defect in the central left pulmonary artery with mass-like features made the diagnostic approach suspicious of a possible tumor. Though, in retrospect, the FDG-PET scan findings of non FDG-avid lesions may have favored the diagnosis of a chronic pulmonary embolism.

This case is representative of a striking presentation of a rare entity- unilateral chronic pulmonary embolism. However, her findings made this diagnosis difficult to distinguish from pulmonary artery sarcoma due to the vast overlap in diagnostic modalities between both conditions. Fortunately the patient had successful thromboembolectomy as she underwent a thoracotomy for what was initially believed to be a mass.

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