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Case Report

Normal ventilation/perfusion lung scan in patients with extensive chronic thromboembolism pulmonary hypertension: A case report

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ABSTRACT

Chronic thromboembolism pulmonary hypertension (CTEPH) is a common cause of severe pulmonary hypertension, resulting in significant morbidity and mortality. In patients with unexplained pulmonary hypertension, a ventilation-perfusion (VQ) scan should be considered the initial diagnostic test of choice. VQ scans are widely available with excellent sensitivity, specificity, and diagnostic accuracy. However, the occurrence of a normal VQ scan in the presence of CTEPH is believed to be rare. In fact, the rate of actual false negatives in VQ scans is unknown because pulmonary digital subtraction angiography and computed tomography pulmonary angiography are rarely performed in patients with a normal VQ scan. This study reports a patient with a high clinical likelihood of CTEPH due to a hypercoagulable state, recurrent deep vein thrombosis, and prior history of acute pulmonary embolism with negative VQ scans. He subsequently underwent pulmonary digital subtraction angiography, which revealed bilateral extensive emboli with partial recanalization of the organized thrombus. The patient underwent successful pulmonary endarterectomy, with marked improvement of his symptoms postoperatively.

Case report

A 23-year-old male was transferred to our hospital after having experienced severe shortness of breath for the previous 8 months; it was initially associated with mild to moderate physical activity but had progressed to occurring with minimal exertion (New York heart functional class 4). He had a history of recurrent deep vein thrombosis (DVT) since the age of 14, secondary to a hypercoagulable state due to antiphospholipid syndrome, which was treated with anticoagulation. There were no symptoms of fever, chest pain, or hemoptysis. His past medical history included acute pulmonary embolism (PE), which was diagnosed approximately 1 year previous after

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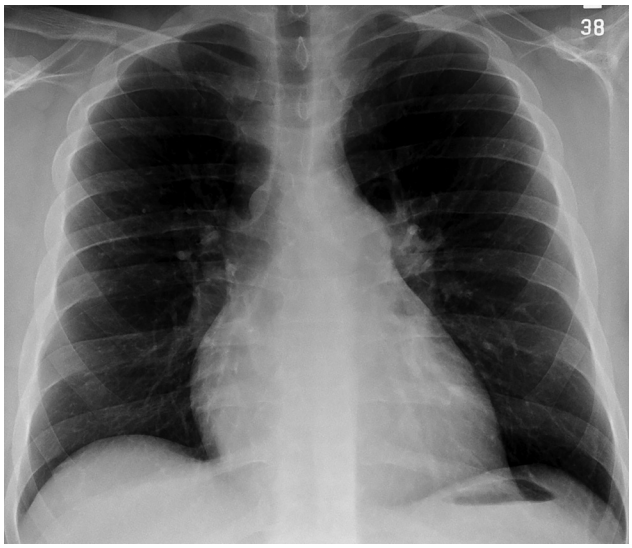

Fig. 1 – Normal chest x-ray with no infiltrate or plural effusion and normal pulmonary vasculature.

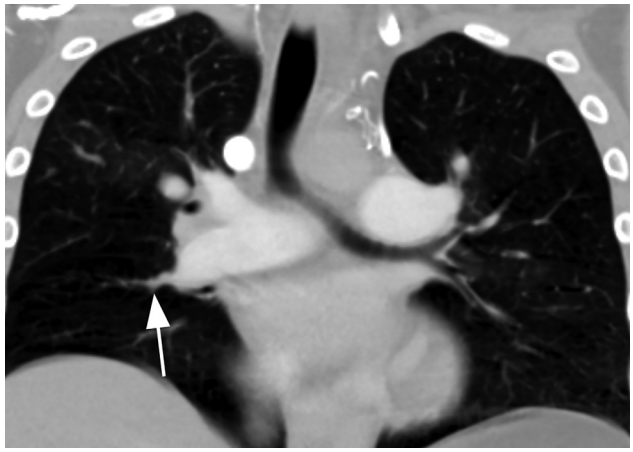

Fig. 3 – Coronal maximum intensity projection reconstruction of a computed tomography pulmonary angiography revealed abrupt caliber change and beaded appearance in the lateral segmental branch of right lower lobe pulmonary artery (arrow).

a syncopal attack, and computed tomography pulmonary angiography (CTPA), which was treated with anticoagulation and resulted in complete resolution of his symptoms. He transferred to our hospital for further treatment of chronic thromboembolism pulmonary hypertension (CTEPH) due to progressive shortness of breath.

On physical examination, the patient appeared to have no signs of respiratory distress. His blood pressure was 125/76, heart rate was 86 beats per minute, temperature was 36.8°C, and respiratory rate was 17 breaths per minute. His cardiac examination showed normal heart sound and was unremarkable for murmur. The respiratory examination revealed equal breath sound with standard percussion, and further tests were normal, with no evidence of acute DVT. Routine blood cells and count were unremarkable, the INR was 2.3, and he was receiving chronic warfarin therapy. Chest x-rays were normal, with no infiltrate or effusion (Fig. 1).

Echocardiography showed 40%-45% mild impairment of the left ventricular function, severe dilation and moderately impaired function in the right ventricle (RV), severe dilation of the right atrial, dilated noncompressible inferior vena cava with inspiration, and moderate to severe tricuspid regurgitation. RV pressure was 80 mm/Hg. A VQ scan was performed without noting typical patterns of CTEPH (Fig. 2). Subsequently, CT pulmonary angiogram was ordered and showed a markedly enlarged RV and markedly dilated main pulmonary artery and pulmonary branch. Abrupt caliber change in the bilateral lower lobe segmental branches was highly suggestive of CTEPH (Fig. 3). Pulmonary digital subtraction angiography (DSA) raveled in the right lung. There was focal narrowing in

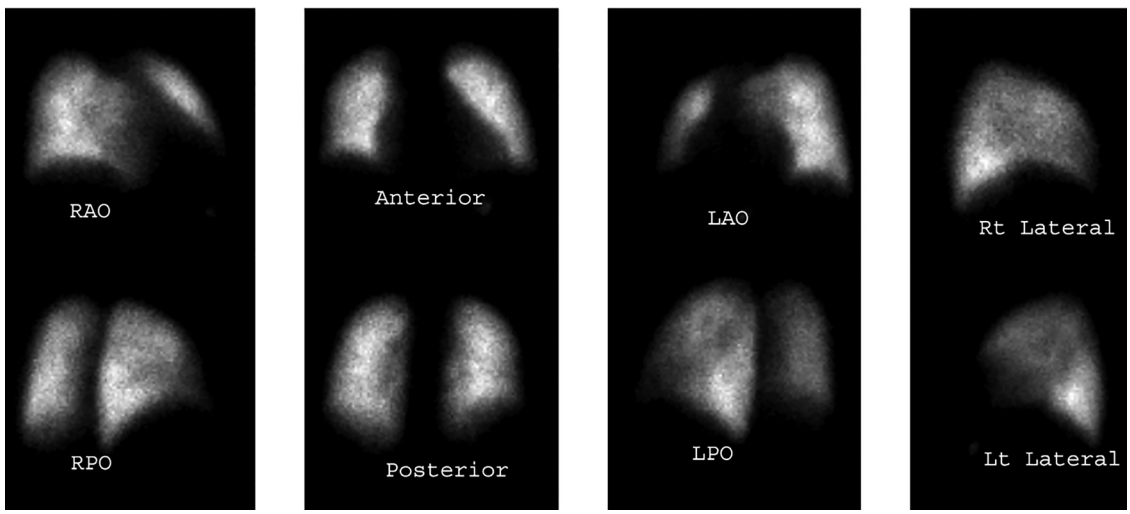

Fig. 2 – Eight-view of perfusion lung scintigraphy demonstrating normal perfusion without segmental or subsegmental perfusion abnormalities; RPO = right posterior oblique; LPO = left posterior oblique; RAO = right anterior oblique; LAO = left anterior oblique.

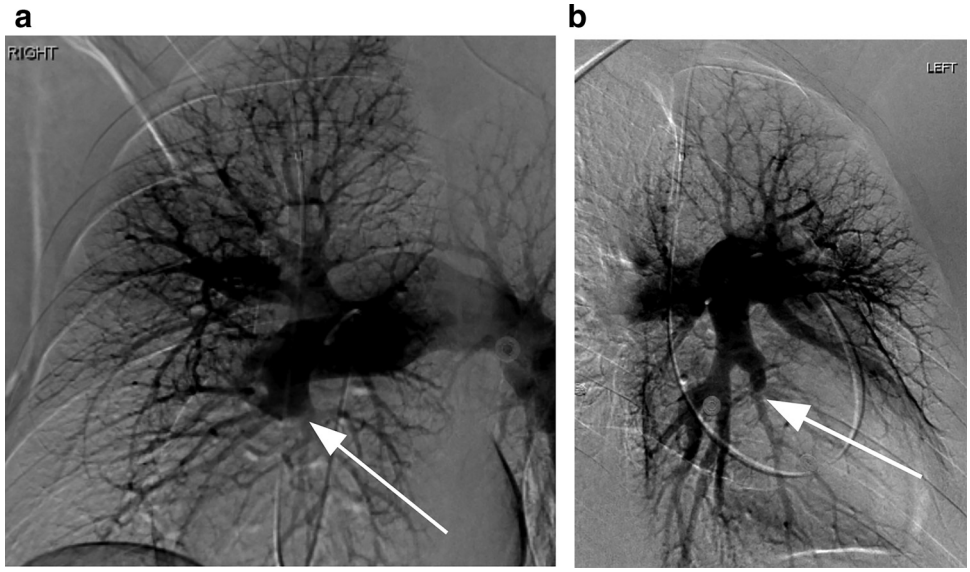

Fig. 4 – (A) Digital subtraction angiography of right pulmonary circulation demonstrating abrupt cutoff of the right lower lobe pulmonary artery, with recanalization of multiple segmental arteries (arrow). (B) Digital subtraction angiography of left pulmonary circulation demonstrates area of narrowing and caliber-size change in left anterior segmental artery of the left lower lobe pulmonary artery (arrow).

the upper posterior segmental branch, with abrupt caliber size change, and the upper posterior and lower medial segments were less vascularized compared to remaining segments (Fig. 4A).

In the left lung, there was focal narrowing in the upper anterior segmental branch, with abrupt caliber size change in the lower medial branch. The inferior inguinal segment was less vascularized compared to remaining segments (Fig. 4B). Pulmonary endarterectomy was performed and revealed that all segmental branches in both the right and left lung were nearly occluded, and there were recanalized channels through the organized thrombus. Layers for endarterectomy were reached, and the patient tolerated the procedure without perioperative complications. A postprocedure echocardiography was done and revealed an estimated RV systolic pressure of 30 mm/Hg. The patient was discharged for continued follow-up with the pulmonary service.

Discussion

A VQ scan is the examination of choice in evaluating for CTEPH from other causes of pulmonary hypertension (PH). According to the most recent guidelines on PH, after a positive VQ scan for CTEPH, invasive pulmonary angiography and CPTA can be used for a detailed workup of the pulmonary arteries [1]. The algorithm recommended by the American College of Cardiology working group suggests VQ scans in all patients with unexplained PH, primarily to assess for CTEPH [2]. A normal VQ scan in the presence of extensive PE, as reported in this case, is rare. Hartmann et al reviewed the literature from 1996 to 2000, and they found only 14 cases of angiographically or autopsy proven PE with normal a VQ scan [3]. However, as pulmonary DSAs or CTPAs are not routinely performed after normal VQ scans, there may have been more cases than are currently known. Tunariu et al. reported a VQ sensitivity of 96%-97.4% and specificity of 90%-95% in detecting CTEPH, vs 51% sensitivity and 99% specificity for CTPA.

In this study, 2 cases that were reported as low probability and did not suggest CTEPH were confirmed by DSA. The final diagnosis was made by CTPA, which showed calcific thrombus in one patient and narrowed but patent pulmonary arterial branch in the other patient [4]. However, a study by Worsley et al reported that among 75 patients, the sensitivity of VQ scans for detecting CTEPH was 100% [5]. In a similar published study, a total of 114 patients suspected of having CTEPH were prospectively enrolled. Patients were classified into 2 categories based on VQ scan interpretation. For threshold 1, high probability and intermediate probability VQ scans were considered positive, and low probability/normal scans were negative. For threshold 2, only high probability VQ scans were considered to be positive, and intermediate to low probability/normal scans were considered to be negative. The sensitivity, specificity, and accuracy of the VQ scans were 100%, 93.7%, and 96.5%, respectively, with threshold 1, and 96.1%, 95.2%, and 95.6%, respectively, with threshold 2 [6].

Normal VQ scans with angiographically proven pulmonary embolisms have been reported in few cases in the literature, with several proposed explanations. The first and most important explanation is that most PE occurring in major pulmonary vessels is nontotally obstructive [7]. Also, prior studies revealed that a narrowing of the lobar vessels by up to 50% could be induced in canine studies without causing marked alteration in pulmonary blood flow [8,9], because the distribution of the radiotracer depicting the lung perfusion is proportional to the relative pulmonary blood flow. The scan may appear normal when there is no complete obstruction by the thrombus

[3]. Second, when there are multiple small emboli bilaterally, they may appear relatively normal. This phenomenon is well known in stress myocardial perfusion, which may cause the scan to appear normal in patients with triple vessel due to balanced ischemia and relatively normal myocardial perfusion [10]. Third, in the case of CTEPH, with relatively longer durations between the occurrence of PE and VQ imaging, recanalization and partial dissolution of the thrombus may occur, with partial restoration of the blood flow [11]. Fourth, small emboli in the basal segment might cause a perfusion defect that may not be depicted by VQ scans, especially if there is respiratory motion. And finally, small perfusion defects may go undetected due to shine through effect [12].

In conclusion, our patient had a high clinical likelihood for CTEPH. He was known to experience hypercoagulable states, recurrent DVTs, and had a prior history of PE 1 year before his current presentation. The most likely explanation of the normal perfusion in our patient is due to extensive symmetrical bilateral pulmonary emboli that may have produced relatively normal perfusion and partially recanalized channels through the organized thrombus. In such rare clinical circumstances, when there is a normal VQ scan and high probability of CTEPH, CTPA must be considered. Also, VQ single-photon emission computed tomography may have higher diagnostic accuracy compared to planar VQ. In case of a discrepancy between VQ and CTPA, as described in this, DSA is worthy to pursue to diagnose CTEPH.

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