

Photo Quiz

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TANAFFOS 

A 35-Year-Old Man with Dyspnea and Hemoptysis

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WHAT IS YOUR DIAGNOSIS?

A man in his thirties was admitted due to new onset dyspnea, right-sided pleuritic chest pain and non-massive hemoptysis since 4 days before admission. On arrival, he was febrile and tachypneic with normal blood pressure. Bibasilar decreased breath sounds and vocal vibration, prominently in the right lung, and 2cm difference in diameter of the left leg were the remarkable findings.

Blunting of the right costophrenic angle was prominent on chest x-ray. Laboratory analysis revealed normal blood cell count, elevated erythrocyte sedimentation rate (125 mm/hr.) and positive quantitative D-Dimer. Blood biochemistry and coagulation profile and urinalysis were normal.

Anticoagulant was initiated with presumptive diagnosis of pulmonary thromboembolism (PTE) and deep vein thrombosis (DVT). Doppler ultrasonography (DUS) and pulmonary computed tomographic angiography (CTA) were performed. DUS was normal, but right sided pulmonary artery embolus was confirmed with CTA (Figures 1 and 2). Interestingly, DUS revealed DVT in the right popliteal artery. Echocardiography was normal.

Despite anticoagulative therapy, dyspnea progressed and the patient's general condition deteriorated. Pleural fluid analysis showed lymphocyte dominant exudate.

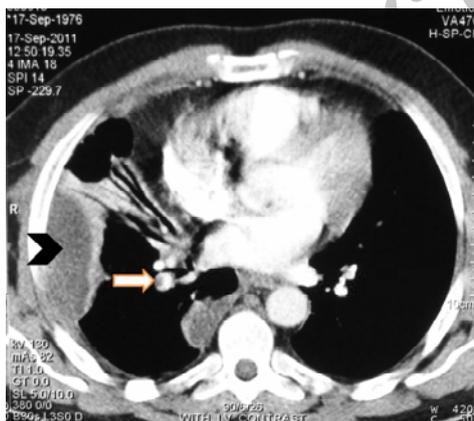


Figure 1. Computed tomography with intravenous contrast revealed filling defect of an embolus in the right division of pulmonary artery (white arrow), loculated pleural effusion (black arrow head) and air-bronchogram in the left lung

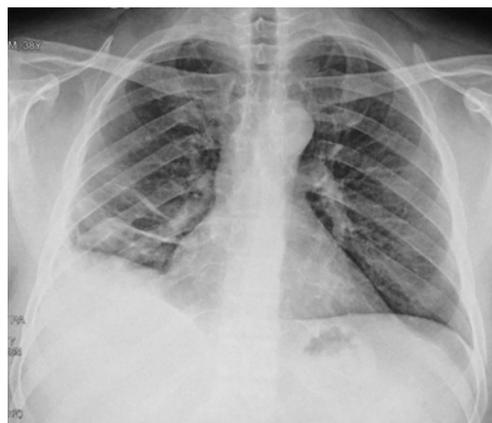


Figure 2. Chest X-ray showed blunting of right costophrenic angle

Answer

Diagnosis: Pulmonary thromboembolism (PTE) due to anti-phospholipid antibody syndrome (APS)

The levels of proteins C, S, V Leiden factor, antinuclear, anticytoplasmic (P and C), and anti-double stranded DNA antibodies were within the normal range. Elevated levels of anti-phospholipid antibodies (anti-cardiolipin antibodies, aCL: IgM and IgG) associated with DVT and PTE confirmed the diagnosis of APS.

Confirmed venous thrombosis, miscarriages or pregnancy morbidities (in females) with positive anti-phospholipid antibodies confirm the diagnosis of APS (1). Frequently, APS occurs secondly to systemic lupus erythematosus (SLE) but may also present as a primary disease (2).

A variety of pulmonary diseases are seen in antiphospholipid antibody syndrome, with the most common manifestations being PTE and pulmonary hypertension (3).

Thromboembolism of lung arteries, pulmonary hypertension, adult respiratory distress syndrome (ARDS), intra-alveolar hemorrhages, postpartum syndrome, and etc. comprise a syndrome named "antiphospholipid lung syndrome" (4).

Pulmonary manifestations may also result from left-sided heart failure due to valvular insufficiency, myocardial infarction or cardiomyopathy. APS is probably involved in the occurrence of some cases of ARDS (2,5). ARDS associated with APS is rare with unclear mechanism. Espinosa and co-workers reviewed 27 patients with this complication which was found to be due to catastrophic APS with a high mortality rate (52%) in the majority of cases (6).

Although recurrent DVTs are the main cause of PTE, it may also be due to in situ thrombosis in pulmonary micro vessels (7). On the other hand, some patients present with

widespread thrombotic occlusions in small pulmonary arteries or alveolar capillary lumens. In a report by Espinosa et al. three of six patients showed isolated microthrombosis on lung biopsy without evidence of pulmonary capillaritis or alveolar hemorrhage (6).

Patients with DVT and PTE should be treated with anticoagulants including heparin followed by warfarin. Lifelong oral anticoagulation is the optimum prophylactic treatment for recurrent thrombosis (6). Furthermore, some thromboembolic events are massive and it seems despite its risk, that pulmonary thromboendarterectomy represents a treatment option for this otherwise lethal condition (7-9).

Nguyen et al. revealed that pulmonary APS has indistinguishable radiological features from pulmonary SLE, suggesting a mutual pathogenetic mechanism. Thus, they claim that probably some of the reported lupus pneumonitis cases in the past might be manifestations of APS rather than SLE (10).

In aforementioned case, the diagnosis of APS and progressive symptoms urged us to start steroids in addition to therapeutic anticoagulants. A few days after administering the combination of enoxaparin and prednisolone, the patient became symptom free and showed remarkable radiologic improvement.

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