Pulmonary embolism: 'the great masquerader' of pneumonia in a patient with progressive supranuclear palsy

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Patients with Parkinson's disease are at risk of developing aspiration pneumonia. Pulmonary embolism is a rare but life-threatening complication in such patients, but could the same be true in progressive supranuclear palsy, an atypical form of Parkinsonism? This case report aims at highlighting the development of unprovoked pulmonary embolism in a patient with progressive supranuclear palsy and also describes how pulmonary embolism can mimic pneumonia in such patients.

Keywords:
Parkinson's disease, progressive supranuclear palsy, pulmonary embolism

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Introduction

Progressive supranuclear palsy (PSP) is an idiopathic neurodegenerative disorder characterized by akinetic rigid Parkinsonism, unsteadiness, dizziness, slowness, falls and pseudobulbar dysarthria. It is sporadic in nature [1]. This unusual syndrome was first described by John Clifford Richardson in 1963, along with John Steele and Jerzy Olszewski, which eventually led to the naming of this syndrome as Steele–Richardson–Olszewski syndrome [2].

Previous studies have suggested that patients with Parkinson's disease could possibly be at risk for developing deep vein thrombosis (DVT) and pulmonary embolism (PE). This case report attempts to explain that patients with PSP are also at risk of developing PE and how PE may mimic pneumonia in such patients.

Case history

Our patient is a 68-year-old man, a retired office worker, with a medical history of PSP who is on synpoda 110 mg (levodopa 100+carbidopa 10 mg) for the past 4 months. He presented to the Emergency Department with a history of fever, mild cough with scanty white expectoration and dyspnea of 1-day duration. He had no other local or systemic complaints. He was treated for left lower lobe pneumonia with intravenous antibiotics 1 month ago. The patient was also a hypertensive on regular medications (losartan and amlodipine).

On examination, the patient was conscious and oriented. His vitals showed a heart rate of 120 beats/min, blood pressure of 90/60 mmHg, temperature of 100°F and respiratory rate of 32 breaths/min with 88% oxygen saturation in room air. Neurological examination indicated bradykinesia, tremors and slurred monotonous speech. The motor system showed cogwheel rigidity and cranial nerve examination indicated defective downward gaze. His respiratory system examination showed fine crepitations over the right infrascapular region. No cardiovascular or abdominal abnormalities were noted. On the basis of the history and clinical findings, an initial diagnosis of right-sided pneumonia with hypotension was made.

However, his ECG showed the presence of the S wave in lead I, and an inverted T wave in III and V1–V4 (Fig. 1). His troponin I was weakly positive. Chest radiography showed features suggestive of right-sided PE, that is, wedge-shaped opacity, enlarged pulmonary artery, horizontal linear opacities in the lower zone and oligaemia of the lung field (Fig. 2). Echocardiogram showed dilatation of the right atrium and ventricle, with mild tricuspid regurgitation and mild pulmonary artery hypertension. Arterial blood gas reported a pH of 7.49, and pO_2 and pCO_2 of 58 and 23 mmHg, respectively. His D-dimer value was elevated (1439 ng/ml). Computed tomography of the chest with intravenous contrast showed submassive embolism of the right and left pulmonary arteries (Figs. 3 and 4). Doppler ultrasound of the bilateral lower limbs did not show any evidence of DVT. His complete blood count, liver, renal and metabolic parameters did not show any abnormalities. His sputum culture showed mild growth of *Moraxella catarrhalis*. Other blood investigations such as protein C and S, homocysteine

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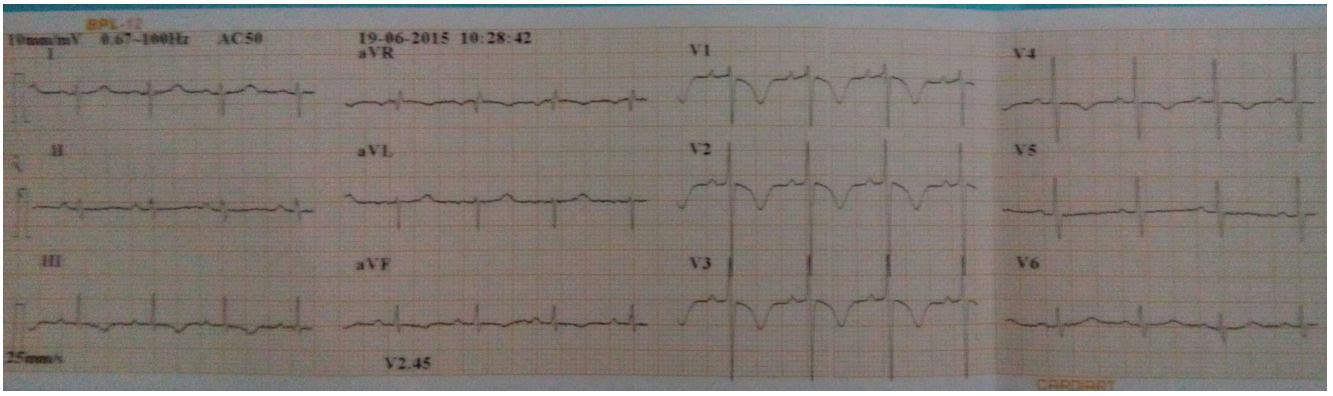
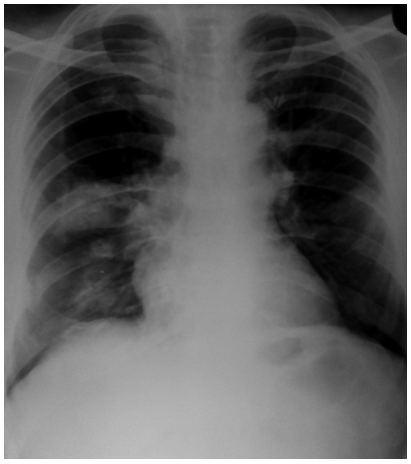
Figure 1



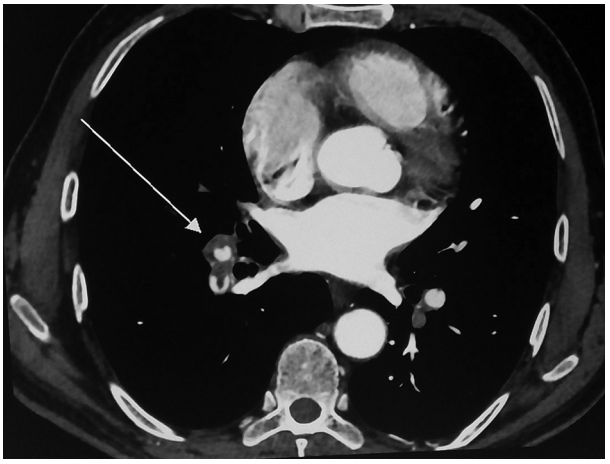
S1Q3T3 pattern with T wave inversion in V1–V4.

Figure 2



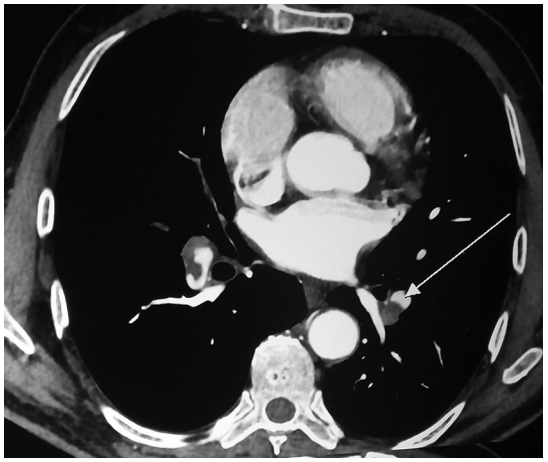
Wedge-shaped opacity, enlarged pulmonary artery, horizontal linear opacities in the lower zone and oligaemia of the lung field.

Figure 3



Right pulmonary artery embolism.

Figure 4



Left pulmonary artery embolism.

levels, antinuclear antibodies and antiphospholipid antibodies were normal.

The patient was started on a weight-based dose of subcutaneous fondaparinux once daily for bridging over to warfarin, which was started at an evening dose of 5 mg. He was also administered levofloxacin as per sputum culture and sensitivity. Following 2 days of therapy, the patient improved symptomatically. He was discharged once the target international normalized ratio (INR) of 2 was achieved.

As this was a case of unprovoked PE, warfarin was continued for life and a target INR between 2 and 3 was maintained. The patient has been attending regular follow-up for his INR values. He was also asked to stop the drug syndopa.

Discussion

PSP is not a rare disease. The estimated annual incidence is about 0.3–1.1 cases per 100 000 persons [3]. It affects both sexes, with a slight male

preponderance. Genetically, these patients frequently show the H1 halotype and the H1/H1 genotype [4]. It is characterized by hyperphosphorylated tau protein aggregates. Macroscopic examination of the brain shows midbrain atrophy and substantia nigra depigmentation, sometimes with mild frontal lobe atrophy, whereas microscopic examination shows neuronal loss, gliosis, neurofibrillary tangles and neuropil threads in the basal ganglia and brainstem [5]. The onset is typically in the sixth decade (range 45 to 75 years) [6]. Clinically, vertical gaze palsy, axial rigidity, pseudobulbar palsy and cognitive impairment have been described as the hallmarks of PSP [7]. Since 1964, a history of fall within the first year of illness has also been included in the criteria [8]. The most common early symptom is an unsteady gait and unexplained falling without loss of consciousness. There is no gait ataxia, but patients tend to lean and fall backward. There is also gradual stiffening and extension of the neck. The patient's face acquires a staring, worried look with a furrowed brow. The face becomes less expressive, speech is slurred, the mouth tends to be held open and swallowing becomes difficult. Many patients may complain of sleep disturbances [6].

PSP becomes progressively worse, but is not directly life-threatening in itself. The most common complications are pneumonia, head injury and fractures caused by falls. There is currently no effective treatment for PSP. In some individuals, the slowness, stiffness and balance problems of PSP may respond to anti-Parkinsonism agents such as levodopa, but the effect is usually temporary. Zolpidem has been found to ameliorate bradykinesia and rigidity. Weighted walking aids have been used to prevent a backward fall. Special glasses called prisms are sometimes prescribed as a remedy for the difficulty in looking down. A gastrostomy may be necessary to tackle swallowing disturbances. Autologous adipose tissue-derived mesenchymal stem cell transplantation show promising results in delaying the progression of PSP [9].

Venous thromboembolism includes DVT and PE. The common predisposing factors include cancer, hypertension, obesity, cigarette smoking, chronic obstructive pulmonary disease, long-haul air travel, oral contraceptive pills, pregnancy, postmenopausal hormone replacement, surgery and trauma. Deficiencies of protein C and S and antithrombin, hyperhomocysteinaemia and antiphospholipid antibody syndrome are some of the rare causes [10].

Postural abnormalities have been reported as a risk factor for DVT in patients with Parkinson's disease [11]. PE may also develop as an adverse reaction to levodopa therapy [12].

Patients with PSP are at risk of thrombosis probably because of decreased mobility secondary to Parkinsonism or as an adverse effect of levodopa. Aspiration pneumonia is a common complication of PSP. This case emphasizes the fact that patients with PSP are also prone to unprovoked PE and how this complication may mimic pneumonia.

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Conflicts of interest
There are no conflicts of interest.

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