



Adenocarcinoma of the lung presenting as thrombotic thrombocytopenic purpura



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ABSTRACT

Acquired thrombotic thrombocytopenic purpura (TTP) usually presents as severe microangiopathic hemolytic anemia (MAHA) and thrombocytopenia in a previously healthy individual. It occurs in approximately three in one million adults and 1 in 10 million children annually. The incidence is increased in females and blacks. Diagnosing TTP and initiating therapy with plasma exchange is a must to avoid preventable complications. Acquired thrombotic microangiopathy has been linked to collagen vascular diseases, use of certain medications, organ transplants, infections, pregnancy and cancer. We report a rare case of a 56-year-old African American male diagnosed with TTP who found to have an asymptomatic adenocarcinoma of the lung. Prompt cancer management resulted in completed remission of the thrombotic microangiopathy.

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1. Introduction

Thrombotic microangiopathy may be the first manifestation of solid tumors such as stomach and lung cancer. Thrombotic thrombocytopenic purpura (TTP) includes the pentad of fever, neurologic abnormalities, thrombocytopenia, hemolytic anemia, and renal disease. It has been observed in many different types of cancer and is most commonly seen in gastric adenocarcinoma followed by carcinoma of the breast, colon, and small cell lung carcinoma [1]. We present a case of acute TTP as the first presentation of undiagnosed lung cancer. Prompt recognition and management resulted in a good outcome.

2. Case presentation

A 56-year-old African American male with no significant past medical history and a history of remote smoking was transferred to our center for evaluation and treatment of altered mental status. The patient reports that he did not have recollection of the events surrounding his admission. On presentation, he was alert and oriented, but he was having difficulty answering questions

appropriately. Physical examination including vital signs was normal. His CBC revealed a WBCs of 9900/ μ L; a hemoglobin of 8.8 g/dL with high reticulocytes (5.9%) and platelets count of 16,000/ μ L. Chemistry revealed a mildly elevated creatinine at 1.3 mg/dL, an indirect bilirubin of 1.4 mg/dL and AST of 46 U/L. LDH was elevated at 886 IU/L. Routine chest X-ray revealed a 3 cm left upper lobe lung mass (Fig. 1), a finding confirmed with a non-contrast chest CT scan (Fig. 2).

CT guided biopsy showed a poorly differentiated adenocarcinoma of the lung.

The patient was started on plasmapheresis which resulted in short-lived improvement. Ultimately, he underwent a left upper lobe lobectomy, after which, his platelets count returned to normal and he went into complete remission without the need for further plasmapheresis sessions.

3. Discussion

Pulmonary tumor thrombotic microangiopathy (PTTM) is a rare complication of pulmonary malignancies and this term was first used in 1990 by Von Hervay et al. [2] but it was first defined in 1937 by Brill and Robertson as “subacute cor pulmonale” [3]. Cancer related thrombocytopenia can result either from bone marrow metastasis or as a paraneoplastic process [4].

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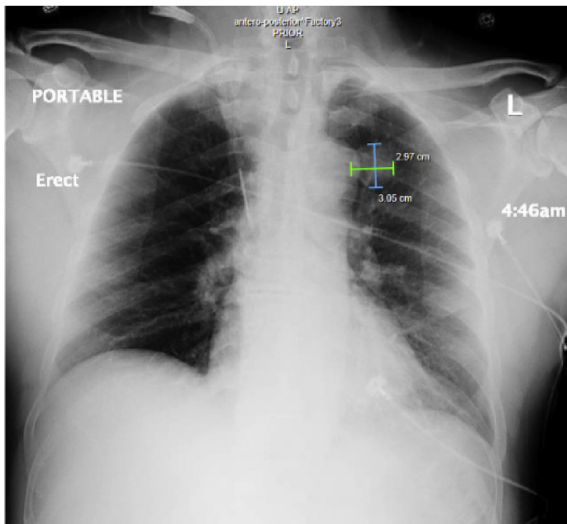


Fig. 1. A chest X-ray showing a 3 cm left upper lobe lung mass.

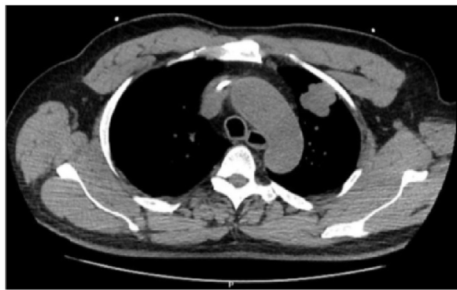


Fig. 2. CT chest showing a 3 cm left upper lobe mass. No mediastinal lymphadenopathy was noted.

TTP is a rare multisystem disorder and affects three patients in one million adults per year [5]. It is a life-threatening disseminated thrombotic microangiopathy. It is predominant in females and in black population [4,6]. Patients rarely present with the pentad of TTP. Recognition of atypical cases and prompt management with either plasmapheresis or treatment of underlying condition is a key factor in improved survival.

According to KRISTIN K. FRANCIS et al. 29 cases have been reported in the literature who presented with TTP-HUS as the presenting symptom before underlying malignancy was diagnosed out of which only three cases had underlying lung malignancy [7]. Patients with underlying malignancy could initially present with microangiopathic hemolytic anemia and thrombocytopenia which can be misdiagnosed as idiopathic TTP (as in our case), leading to inappropriate plasmapheresis and unnecessary delay in the diagnosis and appropriate chemotherapy [8]. A meta-analysis of the literature identified nineteen patients, reported between 1965 and 2005 that were initially suspected for TTP or Hemolytic uremic Syndrome (HUS) and subsequently systemic malignancy was discovered. Fourteen different malignant disorders were eventually found in twenty nine patients to be associated with TTP and HUS cases which included, breast carcinoma, non-small cell lung cancer,

pancreatic carcinoma, renal carcinoma, myelodysplasia, acute lymphoblastic leukemia, diffuse large B-cell lymphoma, and Kaposi's sarcoma, gastric carcinoma, prostate carcinoma, carcinoma of unknown primary, squamous cell carcinoma, colon carcinoma and multiple endocrine neoplasia type I [7].

The presence of a primary malignancy is an essential concern in the differential diagnosis of TTP, particularly in patients with refractoriness to first-line therapy with plasmapheresis. Such cases warrant further workup and investigation to search for the possible occult malignancy especially with risk factors for underlying malignancies which have been reported with TTP. Patients may have no signs or symptoms of malignancy prior to the presentation as TTP as in our case which happened to be the first presentation. The treatment of the underlying malignancy may be the best therapeutic decision in paraneoplastic TTP given its poor response to standard treatments (as in our case). In our literature review we came across only four cases of lung cancer initially presenting as TTP [7,9].

4. Conclusions

TTP is a medical emergency that is almost always fatal if exchange plasmapheresis is not initiated early. This case report reminds clinicians that TTP may be an atypical presentation of lung cancer and occult malignancy should always be kept in consideration in patients with thrombocytopenia of uncertain etiology.

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Conflicts of interest

None.

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