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Primary pulmonary leiomyosarcoma: a rare malignant lung tumor

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Abstract

Primary Pulmonary Leiomyosarcoma (PPL) is an extremely rare malignant tumor. It has been reported that PPL may originate from the smooth muscle of the pulmonary parenchyma, pulmonary arteries, or bronchi. Patients with PPL may be asymptomatic or present with symptoms similar to those observed in other primary lung tumors. This study reports the case of a 61-year-old man who presented with cough and hemoptysis as his primary complaints. Imaging revealed a lung mass, and the patient underwent cryobiopsy. He was subsequently diagnosed with PPL.

Key words: primary pulmonary leiomyosarcoma, malignant lung tumor, case report.

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Introduction

Primary leiomyosarcomas of the lung are extremely rare tumours arising from the smooth muscles of the pulmonary interstitium, bronchi and blood vessels in order of decreasing frequency.[1] Diagnosis is often incidental and based on radiological findings from chest X-ray or Computed Tomography (CT) scans. Due to their rarity and nonspecific symptoms, Primary Pulmonary Leiomyosarcomas (PPLs) are frequently overlooked and misdiagnosed as more common respiratory diseases or lung cancer subtypes. PPLs frequently presents as a round, well-outlined masses on chest radiography. Unlike epithelial tumours, PPL does not exfoliate; hence, bronchoscopy with washings or brushings is usually unrewarding. Pulmonary artery leiomyosarcomas have also been misdiagnosed as chronic thromboembolic disease, as they present with complaints such as dyspnea, chest pain, and features suggestive of right-sided heart failure. In some cases, diagnosis of sarcoma is considered either too late, or not at all. Histological examination is the most reliable way to confirm PPL and its differentiation grade.[2] Preoperative diagnosis of PPL is difficult and surgical resection is often a preferred method for definitive diagnosis and curative treatment. Depending on the size and grade of tumour, chemotherapy, radiotherapy or both may be recommended as an adjuvant therapy.

Case Report

A sixty-one-year-old man presented to our Outpatient Department (OPD) with complaints of cough with yellowish expectoration for past three months, shortness of breath for past one month. There was also history of two episodes of hemoptysis in past one month. Patient had no history of anorexia, chest pain or fever. Patient was vitally stable. Chest examination revealed reduced chest expansion, dullness on percussion on left infrascapular region. Auscultation revealed decreased breath sounds on left infra scapular region and coarse crepitation were heard at the left lung bases. Routine blood investigations were within normal limits. Sputum was negative for acid fast bacilli and malignant cells; sputum culture was sterile.

His chest radiograph showed a uniform left lower zone opacity which was retro cardiac in location (Figure 1). Contrast-Enhanced Computed Tomography (CECT) of thorax suggestive of a soft tissue density mass lesion involving left lung lower lobe (approximately 8.3x6.7 cm). Lesion was encasing the left inferior pulmonary artery and abutting left lower lobe segmental bronchus, with associated collapse consolidation (Figure 2).

On flexible bronchoscopy a whitish growth was visualized at the left lower lobe bronchus protruding out and completely blocking the lumen. Cryobiopsy samples were taken from the growth and sent for histopathological examination (Figure 3).

Histopathological examination of the biopsied samples revealed fibrinous material with bronchial mucosal tissue with hyperplastic mucosal glands. Tumour was made up of hyperplastic spindle cells with hyperchromatic nuclei, which was consistent with pleomorphic sarcoma. Immunohistochemical (IHC) analysis was done, and tumour cells showed strong positivity for Caldesmon, non-specific CK positivity and were negative for S100, CD34, TTF-1, SMA, CK5/6, p63. There was no epithelial

differentiation observed and the overall morphological features favoured a high-grade sarcoma with evidence of smooth muscle differentiation, indicating a leiomyosarcoma. Thus, a provisional diagnosis of PPL was made. Whole body Positron-Emission Tomography (PET-CT) was done which showed metabolically active heterogeneously enhancing soft tissue density mass lesion involving left lung lower lobe. Lesion was seen encasing the left inferior pulmonary artery, mediastinal pleura and descending thoracic aorta. Fluorodeoxyglucose (FDG) uptake was seen in multiple upper, lower paratracheal, anteroposterior window and subcarinal lymph nodes and uptake could not be seen elsewhere in the body therefore a final diagnosis of PPL was made (Figure 4). After a multidisciplinary discussion with a team of cardiothoracic surgeons, radiologists, and thoracic oncologist it was decided to start chemotherapy and then evaluate him for surgery. Patient was started on systemic chemotherapy with doxorubicin and ifosfamide. After two cycles of chemotherapy, patient showed clinico-radiological improvement (Figure 5). Patient was lost to follow up after that and never came for further chemotherapy/follow up.

Discussion

PPL is an extremely rare malignant neoplasm, representing less than 0.5% of all primary pulmonary tumors.[1] These tumors arise from smooth muscle cells of the pulmonary vasculature, bronchi, or interstitial tissues, and their diagnosis is often delayed due to nonspecific clinical presentation. Common symptoms include cough, dyspnea, chest pain, or hemoptysis-features often attributed to more prevalent conditions like tuberculosis, chronic infections or bronchogenic carcinoma.[2,3]

Radiologically, PPL typically presents as a well-defined peripheral or central lung mass, occasionally with evidence of bronchial obstruction or vascular involvement. In our case, the lesion was centrally located and caused complete occlusion of the left lower lobe bronchus, a finding confirmed on bronchoscopy. Given that PPLs do not usually exfoliate, cytological sampling through bronchoalveolar lavage or brushings is often non-diagnostic.[4] Histopathology and Immunohistochemistry (IHC) remain critical for diagnosis. PPLs are composed of spindle-shaped cells with nuclear atypia and mitotic activity, as observed in this patient. Immunohistochemically, PPLs express smooth muscle markers such as caldesmon, desmin, and Smooth Muscle Actin (SMA), while lacking epithelial markers like TTF-1, CK5/6, and p63.[5] In our patient, strong positivity for caldesmon and absence of epithelial markers (*e.g.,* CK5/6, TTF-1, p63) confirmed smooth muscle origin and ruled out carcinoma. The tumor also lacked S100 and CD34 expression, excluding neural or vascular origins respectively. To rule out metastatic sarcoma, whole-body PET-CT is essential. The absence of uptake in extrapulmonary sites confirmed the diagnosis of primary pulmonary leiomyosarcoma in our case, aligning with recent guidelines recommending PET-CT as a standard staging tool.[6] Surgical resection is the mainstay of treatment and offers the best prognosis, particularly in localized disease.[7] However, when tumors are unresectable due to vascular encasement or metastasis, as in our patient, chemotherapy with agents such as doxorubicin and ifosfamide becomes the preferred option.[8] Targeted therapy and immunotherapy have limited roles in PPL, although emerging research is exploring their utility in sarcomas with specific molecular alterations.[9] Unfortunately, the patient was lost to follow-up after two cycles of chemotherapy, underscoring the challenge of maintaining continuity of care in rare and aggressive malignancies.

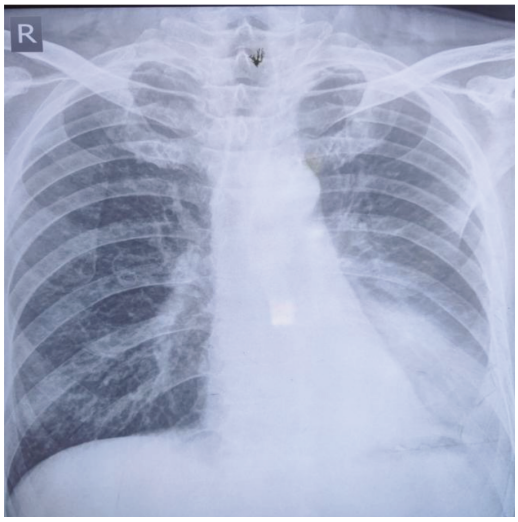

Figure 1. Chest radiograph showed a uniform left lower zone opacity which was retro cardiac in location.

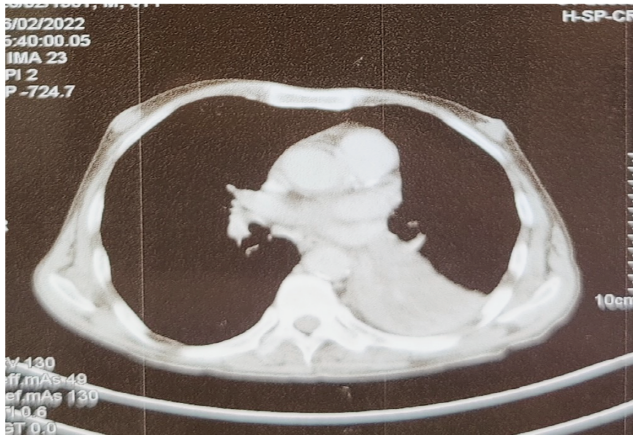

Figure 2. Lesion was encasing the left inferior pulmonary artery and abutting left lower lobe segmental bronchus, with associated collapse consolidation.

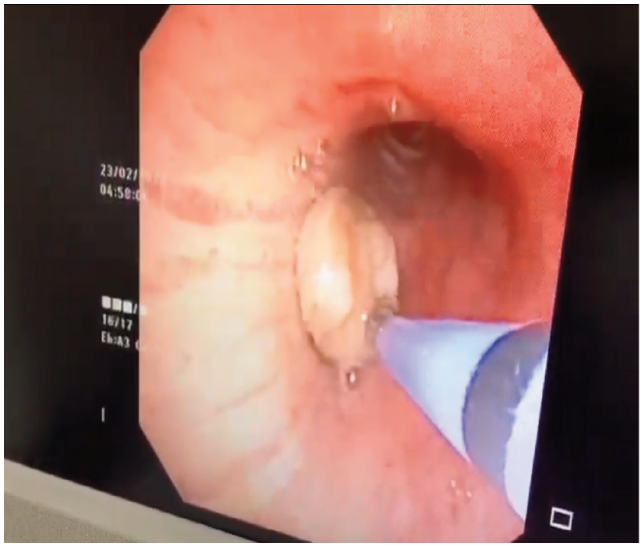

Figure 3. Cryobiopsy samples were taken from the growth and sent for histopathological examination.

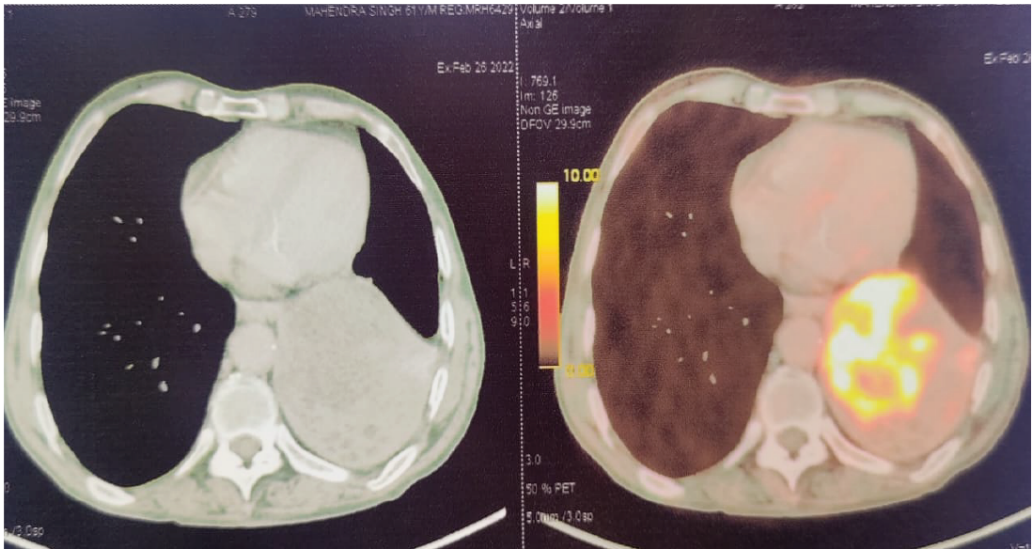

Figure 4. Lesion was seen encasing the left inferior pulmonary artery, mediastinal pleura and descending thoracic aorta. FDG uptake was seen in multiple upper, lower paratracheal, AP window and subcarinal lymph nodes and uptake could not be seen elsewhere in the body therefore a final diagnosis of PPL was made.

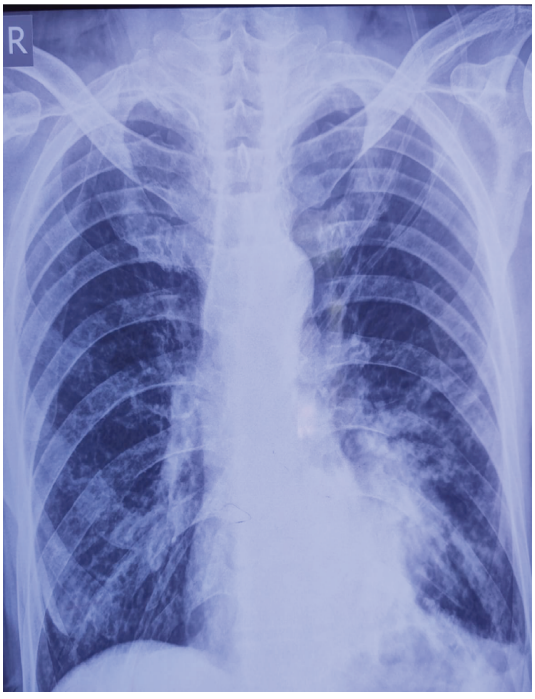

Figure 5. After two cycles of chemotherapy, patient showed clinico-radiological improvement.

Conclusions

PPL is a rare but aggressive malignancy that can present with nonspecific respiratory symptoms, leading to delays in diagnosis. Comprehensive imaging, histopathology, and immunohistochemistry are essential for accurate identification. In unresectable cases, systemic chemotherapy may provide short-term control, although surgery remains the cornerstone of curative management. This case highlights the importance of early multidisciplinary evaluation and the need for ongoing patient follow-up to optimize outcomes in rare thoracic malignancies.

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