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PRIMARY PULMONARY SPINDLE CELL SARCOMA: A RARE CASE REPORT AND LITERATURE REVIEW

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Abstract:
Primary pulmonary spindle cell sarcoma (PPSCS) is an exceptionally rare intrathoracic malignancy with limited cases documented. This report describes a 72-year-old male who presented with persistent cough and weight loss, initially misdiagnosed as pulmonary tuberculosis. Imaging revealed a cavitary mass in the right upper lobe, and bronchoscopic biopsy confirmed a high-grade spindle cell sarcoma of smooth muscle origin. Immunohistochemistry was essential for diagnosis, showing SMA positivity and ruling out epithelial and neural origins. Bronchoscopic intervention achieved partial tumour debulking and symptom relief, followed by systemic chemotherapy with partial radiological response. The absence of distant metastasis and favourable short-term outcome underscore the value of early diagnostic vigilance and multidisciplinary management. This case expands the clinical understanding of PPSCS, especially in TB-endemic regions where misdiagnosis is common. It highlights the critical role of histopathology, immunohistochemistry, and bronchoscopy in diagnosing and managing rare pulmonary tumours.

Keywords: Primary pulmonary sarcoma, spindle cell sarcoma, bronchoscopy, pulmonary mass, immunohistochemistry

Background:

Primary pulmonary sarcomas are rare mesenchymal tumours originating within the lung, without evidence of extrapulmonary primaries. Spindle cell variants consist of poorly differentiated, spindle-shaped malignant cells. Although their aetiology remains unclear, environmental, and genetic factors are suspected. Given that soft tissue sarcomas constitute <1% of malignancies, pulmonary involvement is exceedingly rare and poses significant diagnostic challenges (1, 2).

Primary pulmonary sarcomas comprise only 0.01%–0.05% of lung malignancies (3, 4). Sarcomas typically arise elsewhere, making lung origin rare. Spindle cell variants are even rarer, noted in <4% of cases (5). Most lung sarcomas are secondary; primary origin without metastasis is exceptional. This case exemplifies a rare, de novo pulmonary spindle cell sarcoma. Primary pulmonary sarcomas often mimic common lung diseases, presenting with nonspecific symptoms like cough, haemoptysis, dyspnoea, and chest pain (6, 7). Imaging may reveal masses or cavitary lesions, complicating diagnosis. In this case, persistent haemoptysis and a cavitary lesion led to an initial misdiagnosis of tuberculosis, common in TB-endemic regions (8).

Primary pulmonary spindle cell sarcomas are exceptionally rare, with few cases documented in the literature. A 10-year review identified only five histologically confirmed cases (9). Clinical presentation and outcomes vary, often hinging on early detection and complete surgical resection. Misdiagnosis is common, especially in tuberculosis-endemic regions due to overlapping clinical and radiological features (10, 11).

Definitive diagnosis requires tissue biopsy and immunohistochemical analysis to distinguish sarcomas from histological mimics. Vimentin positivity and lack of epithelial markers support the diagnosis (12). Management typically involves surgical resection, often combined with chemotherapy or radiotherapy. While complete excision offers the most favourable prognosis, recurrence and metastasis exceed 30% despite clear margins (13). The rarity and nonspecific presentation of primary pulmonary sarcomas necessitate heightened clinical suspicion in atypical, treatment-resistant pulmonary lesions. Diagnostic delays are common in regions with high tuberculosis or carcinoma prevalence (14). This case exemplifies a diagnostic shift from suspected infection to confirmed malignancy, emphasizing the critical role of histopathological evaluation.

This case report enhances the limited literature on primary pulmonary spindle cell sarcoma, emphasizing the necessity of histological confirmation in persistent lung lesions. It highlights diagnostic challenges and management strategies, raising awareness of rare thoracic malignancies. The report advocates broader differential diagnoses beyond regionally prevalent conditions in pulmonary clinical practice.

Case presentation:

A 72-year-old male presented to our hospital with a history of persistent dry cough for one month and right-sided stabbing chest pain of similar duration. The chest pain was non-radiating and not associated with exertion. The patient also reported significant unintentional weight loss and loss of appetite during this period. There was no

history of fever, haemoptysis, wheezing, shortness of breath, palpitations, orthopnoea, or paroxysmal nocturnal dyspnoea.

The patient's past medical history was notable for coronary artery disease, for which he had undergone coronary artery bypass grafting two years earlier. He denied any history of hypertension, diabetes mellitus, thyroid disease, pulmonary tuberculosis, or COVID-19 infection, and he had received two doses of the COVID-19 vaccine. His personal history revealed that he had been a heavy smoker since the age of 16 years, consuming approximately 24 bidis per day for 53 years, with a calculated smoking index of 1344. He denied alcohol consumption.

On examination, the patient was stable with a blood pressure of 120/90 mmHg, pulse of 88 beats per minute, respiratory rate of 16 per minute, and oxygen saturation of 96% on room air. Respiratory system examination revealed reduced breath sounds over the right suprascapular, infrascapular, axillary, and clavicular areas. Cardiovascular, abdominal, and neurological examinations were unremarkable.

Baseline laboratory investigations showed mild anaemia (haemoglobin 9.1 g/dL) with a leucocytosis of 12,480/μL, while renal and liver function tests and serum electrolytes were within normal limits. Sputum acid-fast bacillus, cartridge-based nucleic acid amplification test, and cultures showed no growth. A chest radiograph demonstrated a large homogeneous opacity in the right upper lobe suggestive of a space-occupying lesion (Figure 1). A contrast-enhanced computed tomography (CT) scan of the thorax revealed a right upper lobe mass with an abrupt bronchial cut-off sign, associated with post-obstructive changes and pleural involvement (Figure 2).

Subsequent positron emission tomography–computed tomography (PET-CT) confirmed the presence of a large lobulated hypermetabolic mass in the right upper lobe extending into the apical segment, with abrupt cut-off of the apical bronchus and compression of adjacent segmental bronchi. There was evidence of necrosis within the lesion, abutment of the costal and mediastinal pleura, and partial collapse of the right upper lobe. Additionally, mild right pleural effusion with passive atelectasis in the right lower lobe was seen, but there was no evidence of distant metastasis. A few sub-centimetre mediastinal lymph nodes demonstrated mild metabolic activity (Figure 3).

Cardiac evaluation including 2D echocardiography showed an ejection fraction of 55% with grade I diastolic dysfunction, and the patient was deemed fit for invasive procedures. A diagnostic flexible bronchoscopy was performed, which demonstrated an obstructing mass lesion in the right upper lobe bronchus, with an irregular mucosal surface causing narrowing of the lumen (Figure 4). Multiple biopsies were taken from the lesion, and mass debulking was performed using snare excision to restore airway patency (Figure 5). The post-procedural bronchoscopic examination revealed improved visualization of the bronchial lumen, confirming successful tumour removal and clearance of obstruction (Figure 6). Bronchial washings were also collected for microbiological analysis.

Histopathological examination of the biopsy specimens revealed a malignant neoplasm composed of sheets of spindle cells with marked pleomorphism, hyperchromatic nuclei, and brisk mitotic activity, with areas of necrosis and haemorrhage. Immunohistochemistry showed strong cytoplasmic and membranous positivity for smooth muscle actin (SMA) in more than 90% of tumour cells, while markers including P40, TTF-1, Pan-CK, EMA, S100, MyoD1, and CD34 were negative. These findings confirmed the diagnosis of a high-grade spindle cell sarcoma of smooth muscle origin, consistent with primary pulmonary leiomyosarcoma.

The patient subsequently received systemic chemotherapy, and a follow-up chest radiograph demonstrated interval reduction in the right upper lobe mass effect, indicating partial therapeutic response (Figure 7).

DISCUSSION:

Primary pulmonary spindle cell sarcoma is an exceptionally rare malignancy. This case of a 72-year-old male highlights diagnostic challenges, initially mimicking infection, and reinforces the importance of early histopathological evaluation and prompt clinical intervention.

In the current case, the patient presented with persistent cough, weight loss, and a cavitating lung mass, initially raising suspicion for pulmonary tuberculosis. This diagnostic pitfall is consistent with the findings of Huang et al. (2023), who emphasized the frequent misdiagnosis of PPS due to nonspecific symptoms and radiological resemblance to infections, especially in TB-endemic regions (8). Similarly, Qi et al. (2023) and Dermawan et al. (2022) described the diagnostic ambiguity posed by PPS and the importance of histological analysis for definitive diagnosis (7, 11).

Zhang et al. (2024) and Collaud et al. (2020) highlighted that spindle cell sarcomas represent less than 4% of all pulmonary sarcomas and emphasized their diagnostic rarity (5, 15). These tumours usually arise de novo in the lung without a known extrapulmonary source. The current case also demonstrated no evidence of metastasis, consistent with the definition of primary sarcoma. Furthermore, the abrupt bronchial cutoff and necrotic mass seen on imaging mirrors the descriptions by Mohan et al. (2024) and Fiscus et al. (2020), reinforcing the diagnostic clues radiologists and clinicians should be attuned to (2, 4).

Bronchoscopic visualization and debulking performed in this case provided symptomatic relief and tissue access, which aligns with approaches documented by Robinson et al. (2021) and Bushra et al. (2022), who emphasized the importance of endobronchial interventions for diagnosis and palliation in centrally located sarcomas (12, 13). The histological features—marked pleomorphism and spindle morphology—combined with immunohistochemistry positivity for SMA and negativity for epithelial and neural markers were crucial in differentiating this malignancy from mimickers such as sarcomatoid carcinoma and mesothelioma, a diagnostic strategy also supported by Elisa et al. (2022) (16).

Therapeutically, the patient underwent systemic chemotherapy post-bronchoscopic debulking, showing partial radiologic response. This treatment course is supported by Liwei et al. (2020), who observed that multimodal therapy including chemotherapy can improve short-term outcomes even in aggressive sarcomas (14). Robinson et al. (2021) also underscored the benefit of combining surgical and medical management, where feasible, although complete resection remains the gold standard as stated by Zhang et al. (2024) (13, 15). While surgical resection could not be performed in this case due to anatomical and clinical limitations, the choice of chemotherapy alone appears to have stabilized disease progression for the short term.

The current case also contributes a valuable perspective by documenting a rare bronchial obstruction due to spindle cell sarcoma and successful airway restoration via bronchoscopy. As observed by Fiscus et al. (2020), such interventions not only improve quality of life but also enable further therapeutic planning (2). Additionally, PET-CT findings showing localized disease without metastasis in the current patient echo reports by Collaud et al. (2020) and Qi et al. (2023), further confirming that early-stage PPSCS may present without disseminated disease if identified promptly (5, 7).

One of the significant clinical takeaways from this case is the importance of maintaining a broad differential diagnosis for non-resolving pulmonary lesions. This is particularly critical in endemic settings, where infections like tuberculosis are common and may overshadow suspicion for malignancies. The diagnostic delay observed in this case, initially presumed to be TB, mirrors findings by Stefano et al. (2023) and Colin et al. (2022), who reported similar delays in PPS diagnoses (10, 17). Early tissue biopsy and multidisciplinary evaluation could mitigate such delays and improve prognoses.

This case further adds to clinical knowledge by highlighting the critical role of immunohistochemistry in achieving a definitive diagnosis. It also draws attention to the underutilization of molecular profiling in pulmonary sarcomas. Future research should prioritize identifying actionable mutations and developing personalized therapies for these rare thoracic malignancies.

CONCLUSION:

This case highlights the importance of maintaining high clinical suspicion and pursuing early diagnostic intervention in atypical pulmonary lesions unresponsive to standard therapy. Bronchoscopy proved valuable for both diagnosis and palliation, and the favourable response to chemotherapy underscores the role of multidisciplinary care in PPSCS. Histopathology and immunohistochemistry were essential for accurate diagnosis amid misleading clinical and radiologic findings. By documenting a rare pulmonary spindle cell sarcoma mimicking infection, this report adds to limited literature and emphasizes the need for early, comprehensive evaluation. Future research should prioritize molecular insights to guide targeted therapies.

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Figures:

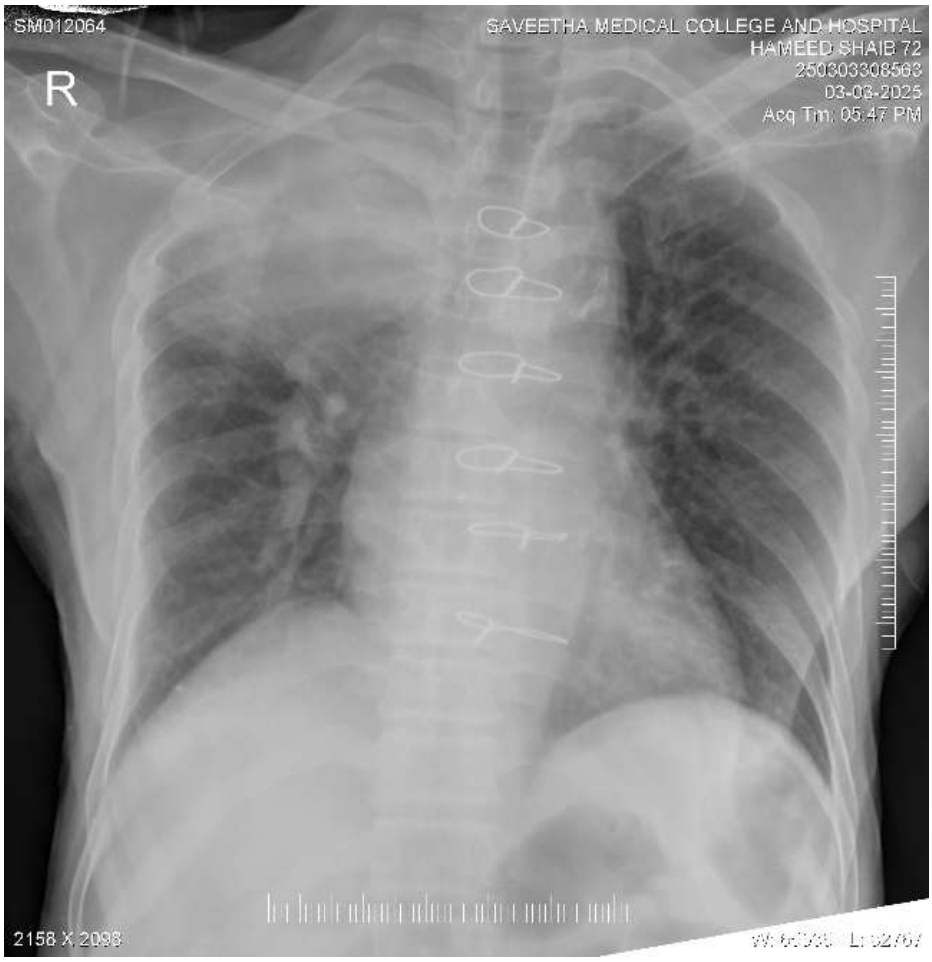

Figure 1: Pre-procedure chest X-ray showing a large homogeneous opacity in the right upper lobe with loss of normal bronchovesicular markings, suggestive of a space-occupying lesion.

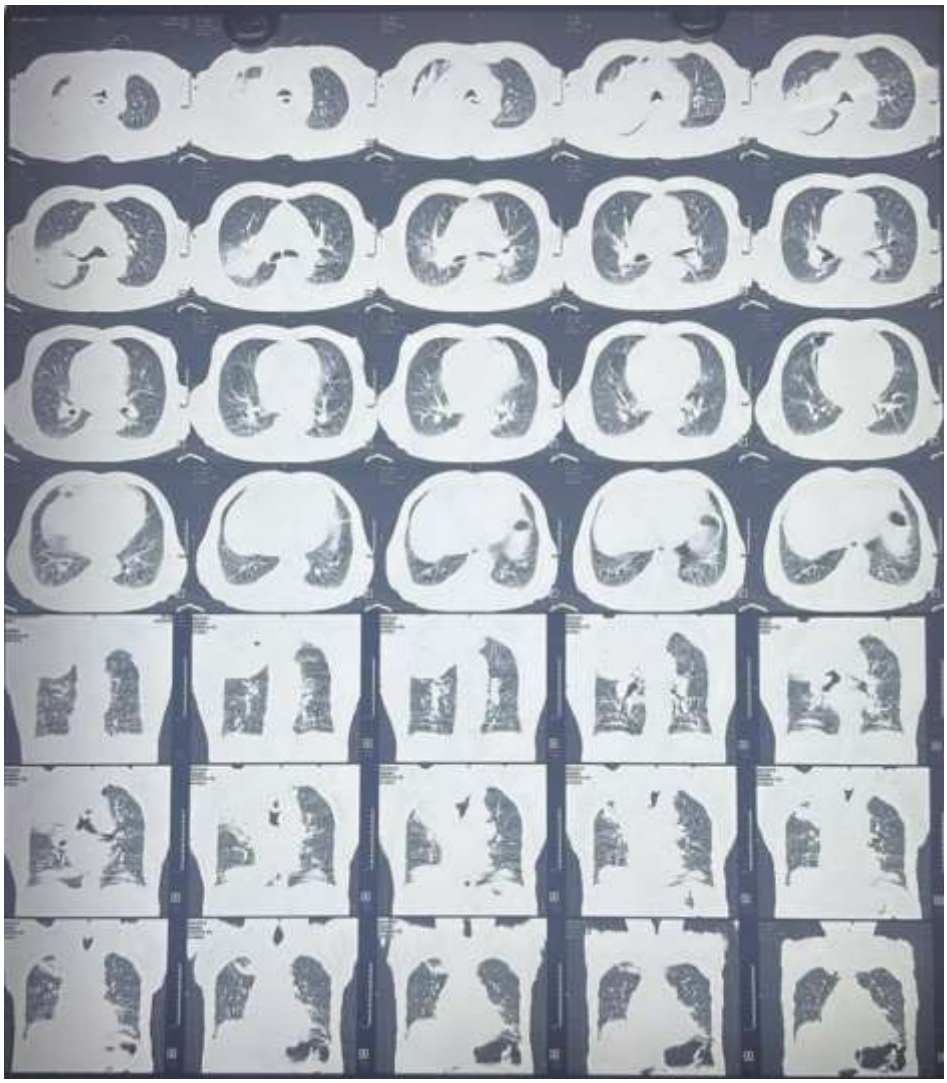

Figure 2: Contrast-enhanced CT thorax demonstrating a large heterogeneous mass in the right upper lobe with bronchial cut-off and associated post-obstructive changes. The lesion appears well-defined with adjacent pleural involvement, consistent with a high-grade spindle cell sarcoma of smooth muscle origin.

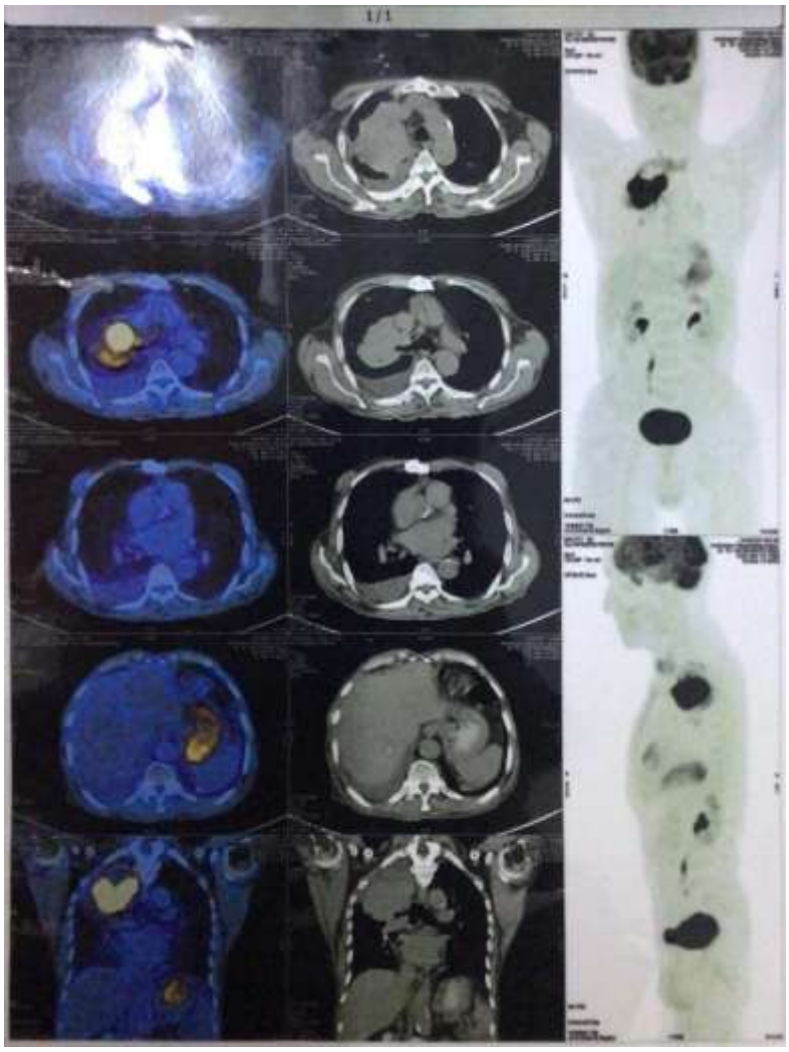

Figure 3: PET-CT scan of the thorax revealing a large hypermetabolic mass in the right upper lobe with abrupt bronchial cut-off and associated pleural effusion, without evidence of distant metastasis.

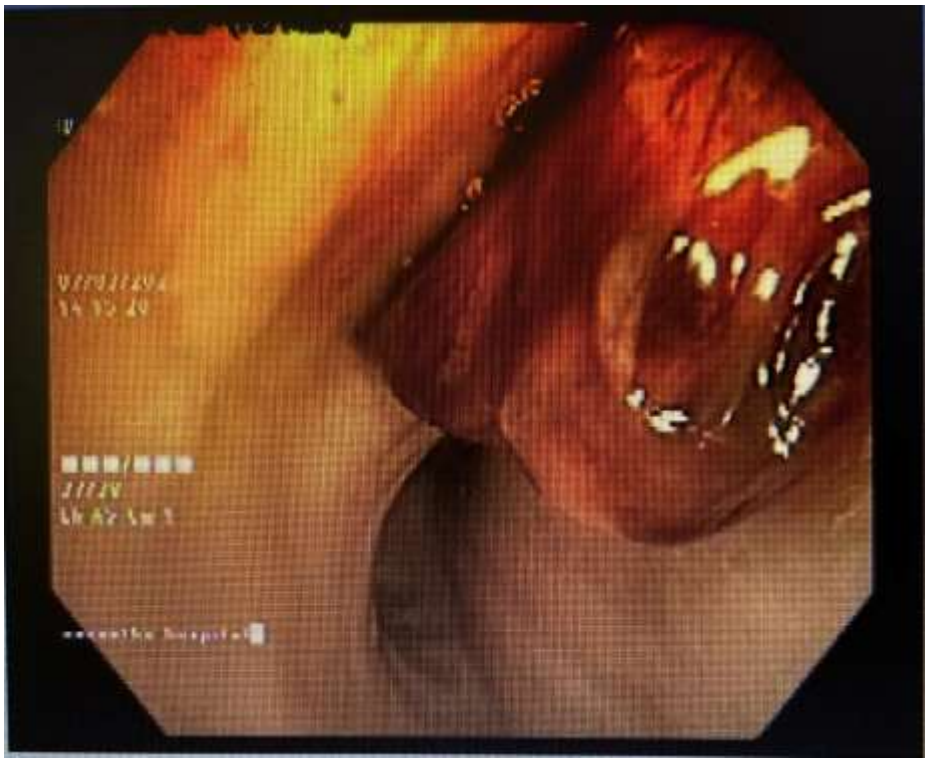

Figure 4: Bronchoscopic view showing a mass lesion obstructing the right upper lobe bronchus, with an irregular mucosal surface and narrowing of the bronchial lumen, consistent with an endobronchial extension of the tumour.

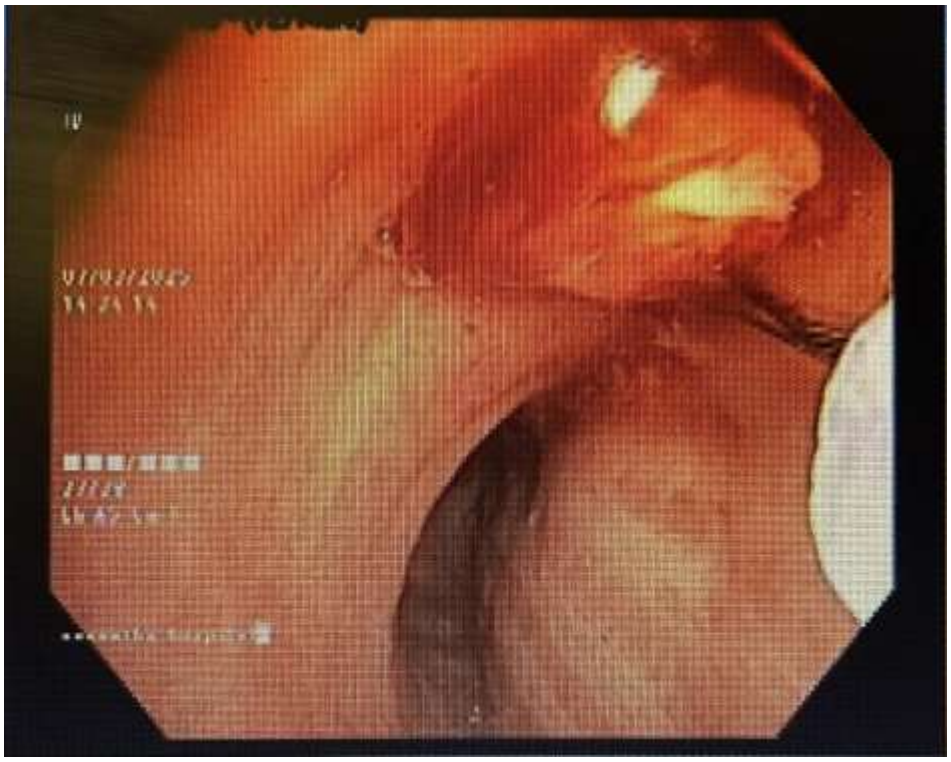

Figure 5: Bronchoscopic view demonstrating snare-assisted removal of the obstructing right upper lobe mass, with partial tumour debulking performed to restore airway patency.

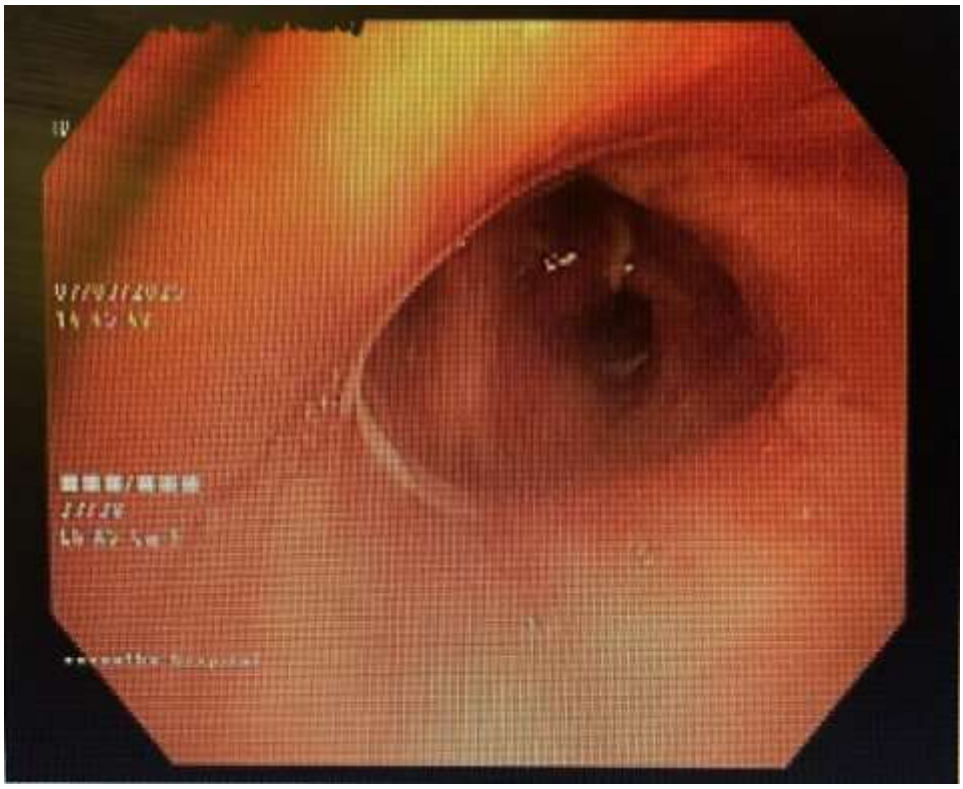

Figure 6: Bronchoscopic view post-debulking of the right upper lobe mass, showing restoration of airway patency with improved visualization of the bronchial lumen after tumour removal.

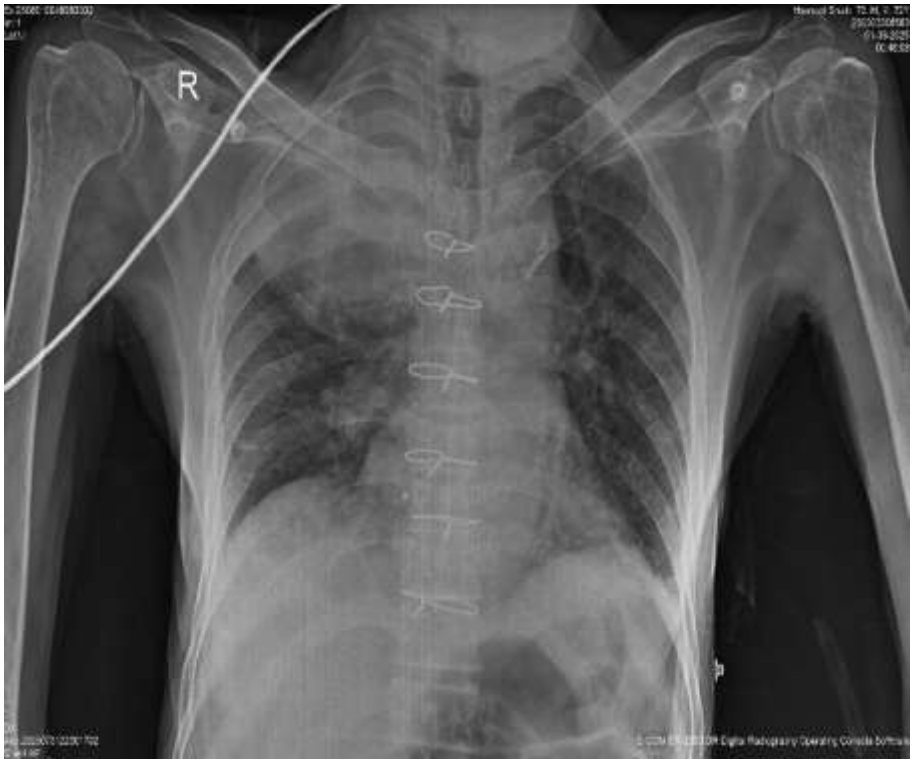

Figure 7: Post-chemotherapy chest X-ray demonstrating interval changes in the right upper lobe following treatment, with reduction in mass effect.

List of Abbreviations

PPSCS	-	Primary Pulmonary Spindle Cell Sarcoma
PPS	-	Primary Pulmonary Sarcoma
TB	-	Tuberculosis

CT	-	Computed Tomography
PET-CT	-	Positron Emission Tomography – Computed Tomography
SMA	-	Smooth Muscle Actin
EMA	-	Epithelial Membrane Antigen
Pan-CK	-	Pan-Cytokeratin
TTF-1	-	Thyroid Transcription Factor-1
P40	-	p40 (ΔNp63) isoform of p63, a tumour marker
CD34	-	Cluster of Differentiation 34 (a cell surface marker)
MyoD1	-	Myogenic Differentiation 1 (muscle-specific transcription factor)
AFB	-	Acid-Fast Bacillus
CBNAAT	-	Cartridge-Based Nucleic Acid Amplification Test
2D Echo	-	Two-Dimensional Echocardiography