RESPIRATORY CASE REPORTS

Pulmonary Carcinosarcoma: A Case Series of Seven Patients and Review of the Literature

Pulmoner Karsinosarkom: Yedi Hastalık Olgu Serisi ve Literatürün Gözden Geçirilmesi

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Abstract

Pulmonary carcinosarcoma is a rare neoplasm of the lung that accounts for 0.3–1% of all primary lung cancer cases, and is characterized by the presence of both carcinomatous epithelial and malignant mesenchymal components. Tumors exhibit aggressive behavior and significant metastatic potential, and there is a high likelihood of recurrence. The primary treatment is surgical, however, the two-year survival rate remains below 10%. We present here an analysis of seven patients diagnosed with lung carcinosarcoma whose diagnosis was confirmed through histopathological examination and the immunohistochemical analysis of tumor biopsies. This case series provides new insights into the clinical characteristics and treatment outcomes of pulmonary carcinosarcoma, highlighting potential differences in a comparison with existing literature.

Keywords: *Pulmonary, carcinosarcoma, cancer, sarkom.*

Öz

Pulmoner karsinosarkom, akciğerin nadir bir neoplazmıdır ve primer akciğer kanseri olgularının %0,3 ila %1'ini oluşturur. Hem karsinomatöz epitel hem de malign mezenkimal bileşenlerin varlığı ile karakterizedir. Bu tümörler, agresif davranış, önemli metastatik potansiyel ve yüksek nüks olasılığı gösterirler. Tedavi öncelikle cerrahidir; ancak iki yıllık sağkalım oranı %10'un altında kalmaktadır. Bu makalede akciğer karsinosarkomu tanısı alan yedi hastanın analizinden elde edilen bulguları sunuyoruz. Tanı, tümör biyopsilerinin histopatolojik incelemesi ve immünohistokimyasal analizi ile doğrulandı. Bu olgu serisi, pulmoner karsinosarkomun klinik özelliklerine ve tedavi sonuçlarına yeni bilgiler ekleyerek, mevcut literatür ile karşılaştırıldığında potansiyel farklılıkları vurgulamaktadır.

Anahtar Kelimeler: *Pulmoner, karsinosarkom, kanser, sarkom.*

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Sarcomatoid carcinoma of the lung (SCC) is an uncommon primary non-small-cell lung cancer variant, five subtypes of which have been identified by the World Health Organization (WHO): carcinosarcomas, giant cell carcinomas, pleomorphic carcinomas, spindle cell carcinomas and pulmonary blastomas (1,2). Carcinosarcoma is a specific subtype of pulmonary sarcomatoid carcinomas that is characterized by high aggressiveness, significant metastatic potential and an elevated risk of recurrence. Diagnosis is established based on histopathological and immunohistochemical findings (3).

Surgery with complete resection is the primary treatment for non-metastatic disease. In contrast, chemotherapy or radiotherapy is preferred for the management of advanced pulmonary carcinosarcoma (2). In the present study we evaluate the treatment responses and prognostic factors of our patient cohort over a two-year period, from January 2022 to January 2024, and compare our findings with existing literature.

CASE

Case 1: A 53-year-old passive smoker presented with left-sided pleuritic chest pain and low-volume hemoptysis of 50 ml per day. The patient's WHO performance status was 0, and oxygen saturation was 97% in ambient air. A comprehensive pleuropulmonary examination revealed normal findings.

Chest computed tomography (CT) identified a small left pleural effusion, and the patient was staged as non-metastatic, with an 80 mm long-axis lesion in the left lower lobe, and as PT4N0 due to the size and extent of the lesion. A fibroscopy biopsy was attempted but was inconclusive. Given the high probability of malignancy and the absence of distant metastases, the decision was made to proceed with a left lower lobectomy and radical mediastinal lymph node dissection. A histopathological evaluation, supplemented by an immunohistochemical (IHC) analysis, confirmed the diagnosis of pulmonary carcinosarcoma. Following surgery, the patient received adjuvant chemotherapy that resulted in favorable outcomes.

Case 2: A 66-year-old chronic smoker with a 40-year smoking history presented with low-volume hemoptysis of less than 50 ml per day and notable weight loss of 6 kg. The patient's WHO performance status was 0, and oxygen saturation (SpO_2) was 96% in ambient air. A comprehensive pleuropulmonary examination revealed normal findings, while chest CT revealed a suspicious 3 cm mass in the left upper lobe. A subsequent positron emission tomography (PET) scan identified a hypermetabolic nodule in the left upper lobe measuring 30*29 mm, with no suspicious active foci detected in the remaining explored areas (Figure 1).

An attempted fibroscopy failed to reach the lesion, and the biopsy results were inconclusive due to a high probability of malignancy. Given the absence of distant metastases, the decision was made to proceed with a left upper lobectomy and radical mediastinal lymph node dissection. The histopathological examination and additional IHC analysis confirmed the diagnosis of pulmonary carcinosarcoma, and the patient was started subsequently on adjuvant chemotherapy, with favorable outcomes.

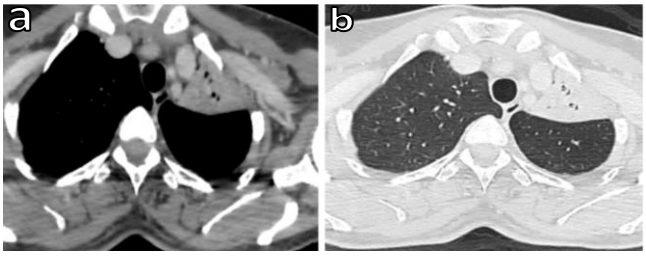

Figure 1: *Thoracic CT scan in mediastinal and parenchymal windows showing a left upper lobar tumor mass*

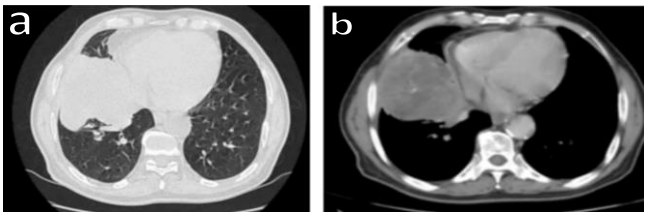

Figure 2: *Thoracic CT scan in parenchymal* ***(a)*** *and mediastinal* ***(b)*** *windows showing a middle lobar tumour mass with regular thickening of the pleural sheets opposite*

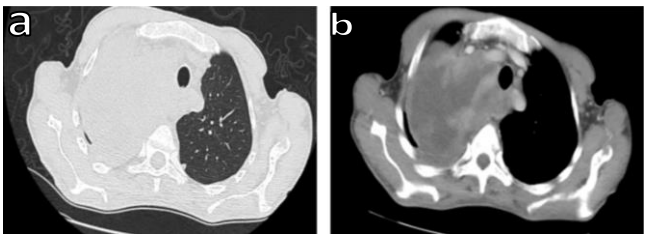

Figure 3: *Thoracic CT scan in the parenchymal window* ***(a)*** *and mediastinal window* ***(b)*** *revealed a right mediastinal-pulmonary tumor mass measuring 120 x 66 mm, in close contact with the trachea, esophagus, and thyroid*

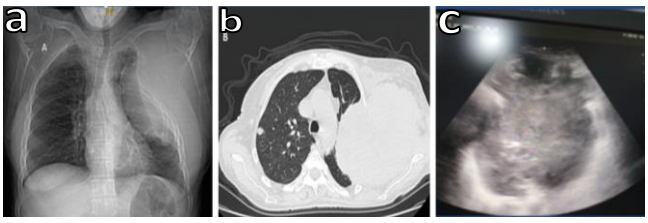

Figure 4: ***(a)*** *Frontal chest X-ray revealed a water-toned opacity occupying the upper two-thirds of the left lung field, with outer margins merging with the chest wall and extending beyond the limits of the thorax, accompanied by costal lysis on the opposite side.* ***(b)*** *Chest CT scan in the parenchymal window displayed a tumoral mass in the left hemithorax, characterized by soft tissue invasion and costal lysis on the opposite side, along with a nodule in the contralateral lung.* ***(c)*** *Ultrasound examination showed a roughly rounded tumor with irregular contours and a heterogeneous echostructure.*

Table 1: Summary Table of Pulmonary Carcinosarcoma Cases

Case Number	Patient Age (years)	Patient Sex	Smoking History	Initial Symptoms	Clinical Findings	Imaging Results	Staging	Diagnosis	Treatment	Surgical Details	Outcome
Case 1	53	Male	Yes	Left-sided pleuritic chest pain, hemoptysis (50 ml/day)	WHO performance status 0, oxygen saturation 97%	Small left pleural effusion; 80 mm lesion	PT4N0	Pulmonary carcinosarcoma	Left lower lobectomy; adjuvant chemotherapy	Radical mediastinal lymph node dissection	Favorable outcomes
Case 2	66	Male	Yes	Hemoptysis (<50 ml/day), weight loss (6 kg)	WHO performance status 0, oxygen saturation 96%	Suspicious 3 cm mass in left upper lobe	PT2N0	Pulmonary carcinosarcoma	Left upper lobectomy; adjuvant chemotherapy	Radical mediastinal lymph node dissection	Favorable outcomes
Case 3	68	Male	Yes	Unexplained weight loss	Mid-lobar mass, lymphadenopathy	CT showed mass with pleural thickening	IVB	Pulmonary carcinosarcoma	Referred for chemotherapy; advanced stage		Passed away prior to treatment
Case 4	63	Male	Yes	Chest pain, dysphonia, dysphagia	WHO performance status 2, oxygen saturation 87%	Right mediastinal-pulmonary tumor mass	IVB	Pulmonary carcinosarcoma	Referred for chemotherapy; advanced stage		Passed away prior to treatment
Case 5	63	Female	No	Chest pain, dyspnea on exertion	WHO performance status 3, large left chest mass	Large heterogeneous mass, rib osteolysis	IVB	Pulmonary carcinosarcoma	Ultrasound-guided biopsy; advanced stage; chemotherapy planned		Passed away prior to treatment
Case 6	61	Male	Yes	Right-sided chest pain	WHO performance status 2, oxygen saturation 96%	Tissue mass in right upper lobe	IVB	Pulmonary carcinosarcoma	Biopsy confirmed diagnosis; planned for chemotherapy		Lost to follow-up
Case 7	58	Male	Yes	Abdominal pain, vomiting, weight loss	Normal clinical examination; pulmonary nodules identified	CT confirmed adrenal incidentaloma	IVB	Pulmonary carcinosarcoma	Initiated on chemotherapy; advanced stage		Succumbed to disease

Case 3: A 68-year-old chronic smoker with a 40-year history of smoking presented without respiratory symptoms. To find the reason for the unexplained weight loss, a chest computed tomography (CT) scan was conducted revealing a mid-lobar mass with regular thickening of the adjacent pleural layers and lymphadenopathy in the Baretey compartment (Figure 2).

Bronchoscopy identified a tumor bud that was obstructing two-thirds of the trachea. A histopathological examination, complemented by additional immunohistochemical (IHC) analysis, confirmed the diagnosis of pulmonary carcinosarcoma. An extension assessment indicated no evidence of distant metastases, and the patient was subsequently referred for chemotherapy. Having been diagnosed at an advanced IVB stage, the patient expired prior to the initiation of treatment, with the cause of death attributed to the advanced stage of the disease.

Case 4: A 63-year-old chronic smoker with a 10-year smoking history presented with chest pain, dysphonia and dysphagia, unexplained weight loss. The patient's WHO performance status was 2 < 50% in bed during the day, with oxygen saturation at 87% in ambient air and signs of central venous congestion. Thoracic CT revealed a right mediastinal-pulmonary tumor mass measuring 120 x 66 mm that was in contact with the trachea and esophagus. There was minimal bilateral pleural effusion, along with secondary pulmonary nodules and metastatic lesions in the axial skeleton (Figure 3). Bronchoscopy revealed complete invasive and budding stenosis of the middle lobe, and a histopathological analysis confirmed pulmonary carcinosarcoma. The patient was referred for chemotherapy, but given his advanced IVB stage he succumbed to the disease prior to the initiation of treatment, with the cause of death linked to the advanced stage of the disease.

Case 5: A 63-year-old female non-smoker with a 10-year history of diabetes presented with chest pain and dyspnea on exertion, coinciding with a deterioration in her overall condition. The patient's WHO performance status was assessed as 3, and a large left-sided chest mass measuring over 7 cm was identified. Computed tomography (CT) revealed a large heterogeneous mass in the lateral aspect of the left hemithorax that extended into the axillary region, with evidence of osteolysis affecting the first four ribs (Figure 4). An ultrasound-guided biopsy confirmed the diagnosis of pulmonary carcinosarcoma, while an extension assessment indicated the presence of metastases in the brain and liver. The patient was subsequently referred for chemotherapy; but expired prior to the initiation of treatment, with the cause of death attributed to the advanced stage of the disease.

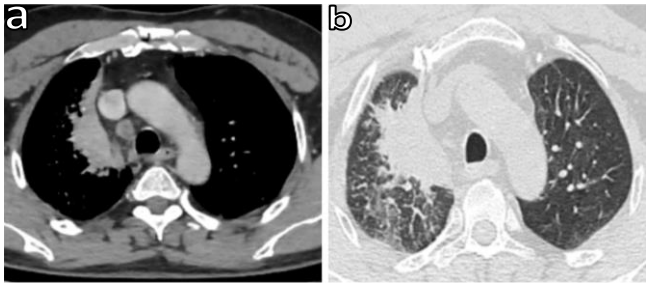

Figure 5: *Thoracic computed tomography (CT) in mediastinal and parenchymal windows reveals a tumor process affecting the ventral segment of the upper right lobe, measuring 8 cm in size. The mass exhibits irregular contours, indicating an aggressive process*

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DISCUSSION

Pulmonary carcinosarcoma is a rare form of tumor. A retrospective cohort study conducted in the United States (SEER: Surveillance, Epidemiology, and End Results) involving 1,052,108 patients diagnosed with malignant tumors of the lung or bronchus reported a prevalence of 0.05% – which is notably lower than the 1% prevalence reported in the present study and may be attributed to differences in sample size (4).

The World Health Organization (WHO) revised the classification criteria for lung cancers in 2004, recommending "sarcomatoid carcinoma" be used as the umbrella term for the range of non-small cell lung cancers with sarcomatous components or that exhibit sarcomatous differentiation (5).

Pulmonary carcinosarcoma is more prevalent in older men in the United States (2), which is consistent with the male/female ratio of 2.5 and a mean age of 61.7±5.25 years identified in the present study.

Clinical signs of pulmonary carcinosarcoma are nonspecific, and depend on the tumor's location. The central endobronchial form typically presents with such symptoms as cough, hemoptysis, dyspnea and recurrent pneumonitis. Conversely, the peripheral form, which is associated

with a poorer prognosis, may present with pain due to the invasion of the chest wall. Additionally, incidental radiological findings have been reported in 6.7% of cases (6). Such tumors predominantly present as peripheral lung masses on CT scans, and are sometimes excavated, invading the pleura and chest wall, or proximal endobronchial or diffuse tumor masses (7). One case in our series had the central form, while the remaining patients exhibited peripheral involvement.

Patients with lung carcinosarcoma do not appear to respond more favorably to standard treatments or to have a better prognosis than those with other types of lung cancer. Complete resection remains the primary treatment for non-metastatic forms of the disease, while for advanced-stage carcinosarcoma, chemotherapy or radiotherapy are often preferred over surgical resection, contributing to intrinsically poor survival outcomes.

Stereotactic body radiation therapy (SBRT) has emerged as an effective treatment modality for inoperable lung carcinosarcoma, with one study even claiming superior survival benefits to conventional fractionated radiotherapy (8). In addition to radiotherapy, chemotherapy has demonstrated some therapeutic efficacy (9).

Targeted treatments with pazopanib have also demonstrated favorable responses in cases of metastatic carcinosarcoma (10). The prognosis for this type of cancer remains poor, with a two-year survival rate of less than 10%, with poorer prognoses associated with tumor size, the presence of distant metastases and the size of the sarcomatous component. Of the seven cases presented here, four succumbed to their illness.

CONCLUSION

Given the rarity of carcinosarcoma, which typically present in adevnced stages, the overall prognosis remains dismal despite advances in treatment modalities. Although the benefits of many of these modalities have yet to be evaluated in large-scale prospective randomized trials, the rarity of the disease suggests such studies are unlikely.

CONFLICTS OF INTEREST

None declared.

AUTHOR CONTRIBUTIONS

Concept - R.A., B.E.S, L.S., M.K.A., M.S., E.M., B.A., S.M., B. A.; Planning and Design - R.A., B.E.S, L.S., M.K.A., M.S., E.M., B.A., S.M., B. A.; Supervision - R.A., B.E.S, L.S., M.K.A., M.S., E.M., B.A., S.M., B. A.; Funding - R.A., B. A.; Materials - R.A., B. A.; Data Collection and/or Processing - R.A.; Analysis and/or Interpretation - R.A., B. A.; Literature Review - R.A..; Writing - R.A.; Critical Review - R.A., B.E.S, L.S., M.K.A., M.S., E.M., B.A., S.M., B. A.

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