

# Pulmonary Spindle Cell Sarcomatoid Carcinoma: A Case Report

## Pulmoner İğ Hücreli Sarkomatoid Karsinom: Olgu Sunumu

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### Abstract

Primary lung sarcomatoid carcinoma (SC) is a rare malignant tumor, with spindle-cell SC representing one subtype of this histological category, only few cases of which have been reported in literature. The treatment and prognosis of pulmonary SC have yet to be clearly determined. Herein we report on the case of a 40-year-old patient with a complaint of chest pain who was found to have a left apical mass in a chest computed tomography (CT). CT-guided lung biopsy and immunohistochemistry confirmed the diagnosis of spindle-cell SC. After radio-chemotherapy, the patient showed an initial partial response and then tumor progression.

**Key words:** Sarcomatoid carcinoma, lung, histology, chemotherapy.

### Özet

Primer akciğer sarkomatoid karsinomu (SC) bir nadir görülen malign tümörlerden olup iğ hücreli SC, bu histolojik kategorinin bir alt türünü temsil eder. Literatürde sadece birkaç olgu bildirilmiştir. Tedavi ve prognoz, pulmoner SC için net olarak belirlenmemiştir. Burada, bilgisayarlı toraks tomografide (BT) sol apikal kitle saptanan ve göğüs ağrısı araştırması yapılan 40 yaşındaki bir hastayı sunuyoruz. BT eşliğinde yapılan akciğer biyopsisi ve immünohistokimya inceleme sonunda hastaya iğ hücreli SC tanısı kondu. Radyo-kemoterapi uygulandı. Başlangıçta kısmi bir cevap alınmakla beraber sonra bir tümör progresyon gösterdi.

**Anahtar Sözcükler:** Sarkomatoid karsinom, akciğer, histoloji, kemoterapi.

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Primary pulmonary sarcomatoid carcinoma (PSC) is a rare form of non-small cell lung cancer (NSCLC) that accounts for less than 0.5% of all lung cancers (1,2). It mainly affects men aged 50–80 with a history of smoking (3). It has been defined as a poorly differentiated non-small cell carcinoma containing a component of sarcoma or sarcoma-like elements whose differentiation is based on immunohistochemical techniques (2). The 2015 World Health Organization (WHO) classification identifies spindle-cell SC as a very rare subtype of SC (2,4), the latter being a tumor with a high grade of malignancy and with a poor prognosis. Treatment options are still not well established (5). We report here on a new case of a locally advanced pulmonary spindle-cell SC.

## CASE

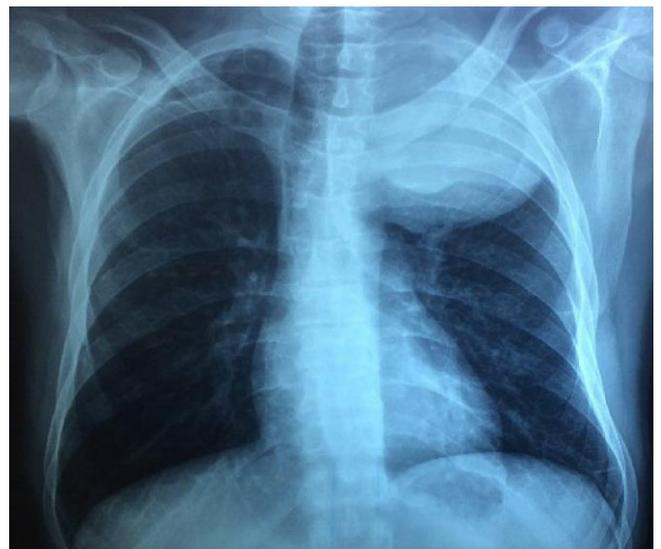
A 40-year-old male patient who was a heavy smoker was explored following a complaint of left apical chest pain that had been evolving over the past four months. He had no prior medical history. A chest X-ray revealed a well-defined opacity in the left upper lung field (Figure 1). A chest computed tomography (CT) revealed a left apical mass measuring 12x8x7 centimeters with some areas of necrosis and vascular contact (Figure 2). The mass was in contact with the neural foramina T3-T4 and T4-T5. There were also multiple mediastinal and subclavian lymph nodes. Brain and abdominal CT showed no evidence of metastasis. Magnetic resonance imaging (MRI) of the T-spine revealed a supraclavicular invasion of the brachial plexus and of the first and second intercostal spaces. Bronchoscopy was normal. A CT-guided lung biopsy was performed, and the histopathological examination revealed a malignant spindle-cell proliferation with focal necrosis. These cells had eosinophilic cytoplasm and an oval nucleus with moderate to marked atypia, as well as several mitotic figures. An immunohistochemical study showed diffuse positivity for vimentin and a focal positivity of some tumor cells for CK7 (Figure 3).

The patient underwent six cycles of chemotherapy involving a combination of cisplatin and docetaxel, as well as curative chest radiation therapy (70 grays). A chest CT scan performed after radio-chemotherapy revealed 30% tumor regression according to RECIST criteria. The patient was followed up regularly. The tumor remained stable with no metastasis until one year later, when local tumor progression was detected in a control chest CT scan. The patient is currently undergoing second line chemotherapy with gemcitabine. He is still alive and is

maintaining a good general condition (performance status =0).

## DISCUSSION

Pulmonary sarcomatoid carcinoma is a rare subtype of NSCLC that accounts for less than 0.5% of all malignant lung tumors (1,6). According to the 2015 World Health Organization (WHO) criteria, sarcomatoid carcinoma can be classified into five subcategories: pleomorphic carcinoma, giant-cell carcinoma, spindle-cell carcinoma, carcinosarcoma and pulmonary blastoma (2). Pulmonary spindle-cell SC is extremely rare (1,2,6), and given the rarity and the difficult diagnosis of this subtype, most of the available information has come from small retrospective studies. Pulmonary SC mostly affects men in their sixth and seventh decades (7). An association with heavy smoking, as seen in our case, has been noted (8). Our patient is younger than the average age noted in literature.



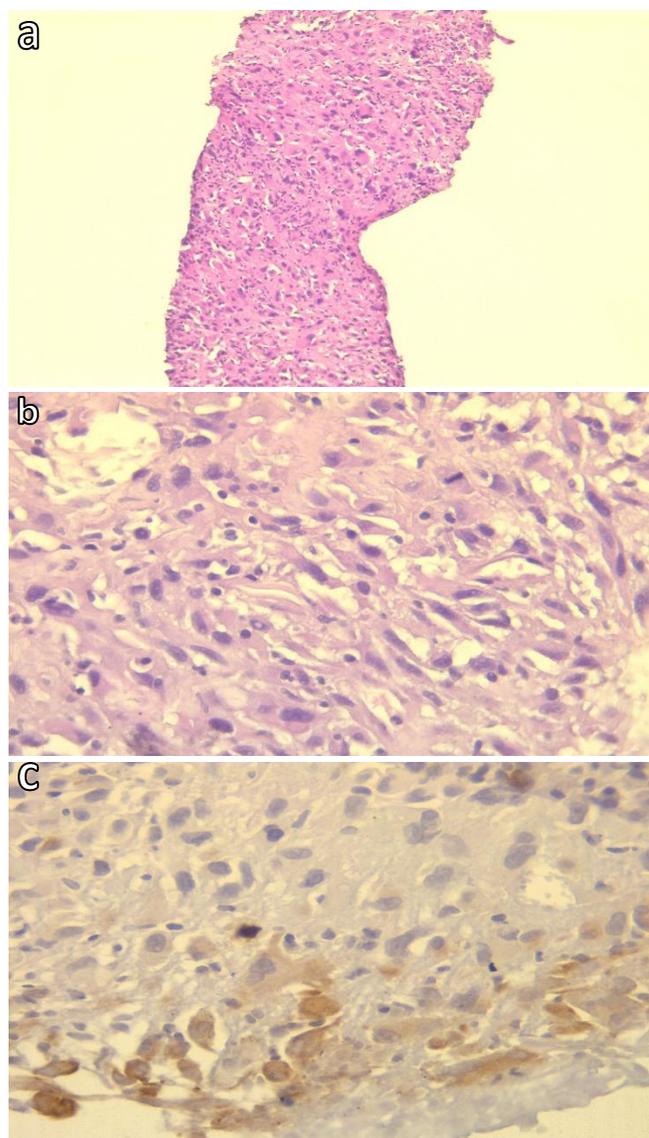
**Figure 1:** Chest X-ray: well-defined mass in the left upper lung field



**Figure 2:** Axial chest CT scan (mediastinal window setting): left apical mass measuring 12x8x7 centimeters with some areas of necrosis, vascular contact and parietal extension

There are no specific symptoms or clinical features differentiating SC from other NSCLCs (8). In some reports, PSC may act aggressively with a more rapid invasion of such adjacent structures as the pleura, chest wall, diaphragm or mediastinum. It is often seen as a large peripheral mass in imaging with a predilection for the upper lobe (3).

PSC is a mixture of epithelial and mesenchymal elements (5). The histogenesis of SC remains unclear, although several hypotheses have been put forward to explain it. A common origin of epithelial and mesenchymal components with an epithelial-mesenchymal transition (EMT) of the original clone represents the most agreed theory in literature (6). The preoperative diagnosis of



**Figure 3:** Histopathology: CT-guided biopsy involved malignant spindle cell proliferation. Low magnification view (HEx40) (a), spindle cells with oval nuclei and eosinophilic cytoplasm, mitoses are numerous. High magnification view (HEx400) (b), Focal positivity of some tumor cells for CK7 at immunohistochemical study (IHCx400) (c)

Pulmonary SC is difficult due to the heterogeneity of the components and the poor cell differentiation (9), and this makes it difficult to identify the exact sub-type of SC from a small biopsy sample. In different studies, the post-operative pathological diagnosis differed from the pre-operative diagnosis of the sub-type of SC (6). Predominantly spindle cell-shaped tumor cells are found in spindle-cell SC (2). An immunohistochemistry (IHC) study is very useful for the diagnosis of PSC. The sarcomatoid component expresses vimentin, whereas the epithelial component may be stained with cytokeratins, an Epithelial Membrane Antigen (EMA) or a carcino-embryonic antigen (CEA) (4). In our case, the diagnosis of spindle-cell PSC was confirmed, since the tumor was mainly composed of spindle-shaped cells with positive staining by vimentin and cytokeratin at IHC.

Given its rarity, pathological heterogeneity and rapid progression, there are no specific recommendations for PSC (5). Early stage surgical treatment represents the standard of care, and the complete surgical removal of the tumor with negative tumor margins is the recommended treatment approach (5,6). Radiation may be effective in addition to surgery if the resection margins show residual tumor, reducing the rate of local recurrence of PSC to about 15% (10). That said, there is no clear protocol of chemotherapy for metastatic or locally advanced disease. The response of PSC to systemic chemotherapy is controversial (3). In some studies, a platinum-based combination regimen was more effective on overall survival than mono-chemotherapy without platinum (11). In our case, a combination of cisplatin and docetaxel reduced the size of the tumor by 30%. No predictive factor indicating response to chemotherapy has been identified (6). There is little data regarding targeted therapy or immunotherapy in PSC (5,12).

The prognosis of PSC is poor in comparison to other types of NSCLC (5). There are several factors affecting prognosis, especially in locally advanced cases of the disease with a tumor size greater than 6 cm, lymph node invasion and the presence of metastasis (6,13). PSC is also characterized by high relapse and low survival rates (6).

## CONCLUSION

Pulmonary sarcomatoid carcinoma is a rare lung cancer, the diagnosis of which remains difficult due to the undifferentiated morphology and the variety of clinical presentations. Surgical resection in the early stages remains the optimum treatment approach, making early diagnosis

essential, although the prognosis remains very poor. The creation of national registries based on new international collaborative studies is needed to better understand the benefits of the different treatment approaches of chemotherapy, targeted therapy or immunotherapy in locally advanced and metastatic forms.

## CONFLICTS OF INTEREST

None declared.

## AUTHOR CONTRIBUTIONS

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

## YAZAR KATKILARI

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