ISSN: 2476-8642 (Print)
ISSN: 2536-6149 (Online)
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Annals of Health Research

VOLUME 8
NO. 1
JAN. - MAR., 2022

IN THIS ISSUE

PUBLISHED BY THE MEDICAL
AND DENTAL CONSULTANTS ASSOCIATION
OF NIGERIA, OOUTH, SAGAMU, NIGERIA.
www.mdcan.oouth.org.ng

Annals of Health Research
Volume 8, Issue No 1: 74-80
March 2022
doi:10.30442/ahr.0801-07-158

CASE REPORT

Spindle Cell Sarcomatoid Carcinoma of the Lungs in a Nigerian Woman: A Case Report

Ojo OT

Department of Medicine, Lagos State University Teaching Hospital, Ikeja, Lagos.

***Correspondence: Dr OT Ojo, Department of Medicine, Lagos State University Teaching Hospital, Ikeja, Lagos.
E-mail: ojofemi911@yahoo.com; ORCID – https://orcid.org/0000-0002-5846-2457.**

Summary

Spindle cell sarcomatoid carcinoma of the lung is a very rare but highly aggressive tumour. The presentation is usually non-specific and can be confused with bronchogenic carcinoma, even on imaging. Tissue biopsy, with histology and immunohistochemistry, is very helpful in diagnosing and differentiating sarcomatoid cell cancer from bronchogenic carcinoma. Surgical resection is the treatment of choice, and if it is not possible, palliative chemotherapy may be an option. This report is about a rare case of spindle cell sarcomatoid sarcoma of the left lung in a flight attendant who presented with left-sided, dull aching chest pain, associated shortness of breath, and cough that was occasionally productive, haemoptysis and weight loss of one-month duration. The tissue biopsy with histology and immunohistochemistry supported the clinical diagnosis of the tumour. Although rare, spindle cell sarcomas may occur in very few patients. The clinical history, imaging and pathologic findings are essential in preventing misdiagnosis of these patients.

Keywords: Giant cell cancer, Lung cancer, Non-small cell cancer, Sarcomatoid cancer, Spindle cell cancer.

Introduction

Spindle cell sarcomatoid carcinoma is a rare, high-grade, poorly differentiated, non-small cell cancer. The incidence of sarcoma is 0.3% to 1.3% of all lung cancers. [1] Spindle cell sarcoma is one of the five histologic variants of sarcomatoid carcinoma, including pleomorphic carcinoma, spindle cell carcinoma, giant cell carcinoma, carcinosarcoma, and pulmonary blastoma. [2-4] Spindle cell carcinoma consists of a pure population of spindle cells, usually involving the periphery of the lung. [5] Many cases have been described in retrospective studies. [5, 6] Endobronchial spindle cell carcinoma is extremely rare, and only a few cases of endobronchial leiomyosarcoma have been reported. [7, 8] However, there has been no report of spindle cell carcinoma from Nigeria. The knowledge of the spindle cell sarcoma as one of the histologic types of non-small cell lung cancers is essential for consideration in the differential diagnosis of patients with lung tumours. The case of a young female, non-smoking patient, with incidental findings of spindle cell carcinoma of the lungs, supported by reports of left-sided pleural effusion and a basal mass, on chest tomographic scan is described in this report.

Case Description

A 36-year old female flight attendant presented with left-sided, dull aching chest pain and weight loss of about 5kg of one-month duration. There was associated occasional shortness of breath, occasional dry cough and a few episodes of haemoptysis of about 50 ml per episode containing clotted blood. The severity of her symptoms progressively worsened. The pain was not relieved by simple analgesics, disturbing her sleep at night. There was no history of fever, wheeze, or anorexia. She was not a cigarette smoker, and there was no history suggestive of secondary exposure to smoking or parental smoking. She had no history of exposure to vapour, gas or chemicals in childhood. She had previously removed a lump from her breast but denied a family history of lung diseases.

On general examination, she was not pale, afebrile with a temperature of 36.7^{o}c and had no finger clubbing or engorged neck vein. She had palpable superficial left axillary lymph nodes. Respiratory system examination revealed a respiratory rate of 24 cycles /minute and oxygen saturation (SPO2) of 95% in room air. There was a shifting of the trachea to the right, reduced movement of the left hemithorax, and reduced tactile fremitus on the left middle and lower lung zones. There was reduced vocal resonance, stony dull percussion note, and reduced breath sounds on the same side. On cardiovascular examination, the pulse rate was 90 beats/minute, blood pressure was 120/80 mmHg, and normal heart sounds with no murmur. Other systemic examinations were essentially normal.

Initial laboratory investigations revealed a full blood count showing neutrophilic leukocytosis with an elevated erythrocyte sedimentation rate (40 mm/hour). Both sputum testing for acid-fast bacilli and Gene Xpert were negative for tuberculosis, and the sputum microscopy and fungal culture were also negative. The Chest X-Ray in the posteroanterior view showed obliteration of the left costophrenic sulcus and hemidiaphragm with a mass overlying the left hemithorax. A chest computer tomographic scan showed left-sided pleural effusion with left lower pleural-based mass (Figure 1).
The patient had diagnostic and therapeutic thoracentesis, and the pleural aspirate analysis was negative for tuberculosis, pyogenic organisms or malignant cells. The cytology shows intense lymphocytes, foamy macrophages and neutrophils in the background of fibrin amid red cells. The aspirate Adenosine Deaminase (ADA) and Lactate dehydrogenase (LDH) were normal. The serum Anti-Nuclear Antibodies

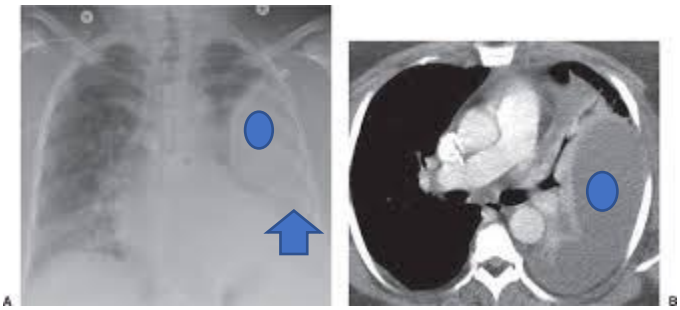


Figure 1: Blue ball identifies the pleural-based mass while the blue arrow shows a pleural effusion
A - Chest X-Ray showing obliteration of the left costophrenic sulcus and hemidiaphragm with a mass overlying the left hemithorax.
B - Chest computer tomographic scan showing left-sided pleural effusion with a pleural based mass.

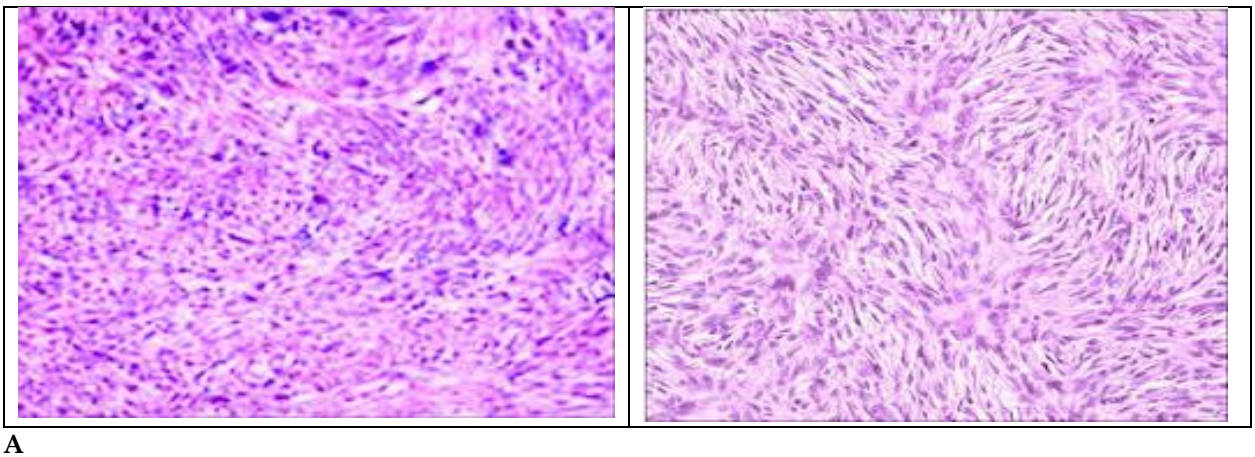
(ANA) was positive; however other screening for connective and inflammatory diseases (Anti-double Stranded DNA Antibodies (anti-dsDNA), Anti Sjogren's syndrome-related antigen A and Anti Sjogren's syndrome-related antigen B (anti-SSA/Ro and SSB La) were negative except for elevated c3 (218mg/dl).

The axillary lymph node biopsy showed expansion of the sinuses packed full with histocytes and lymphocytes and scattered macrophages containing brownish pigments suggestive of reactive lymphadenopathy. The abdominopelvic computerized tomography was essentially normal. The presumptive diagnosis was lung malignancy with a differential diagnosis of systemic lupus erythematosus. A CT-guided core needle biopsy of the lungs was done, and the histology showed features of a malignant mesenchymal tumour.

The sample was sent to a laboratory overseas for immunohistochemistry which showed short plum spindled cells with moderate amounts of eosinophilic cytoplasm and atypical mildly pleomorphic and deeply hyperchromatic nuclei. Few mitoses were observed, and the Ki- 67 index was in the region of 10%. The tumour cells expressed broad spectrum cytokeratins (AE1/3) but were negative with CK5/6. There was a variable weak expression of P63 in a small portion of the tumour cells. The cells were completely negative with P40, myogenin, calretinin, CD34, CD 45, TTF-1, Nasin, synaptophysin, chromogranin and CD 56. These histologic features agreed with the diagnosis of a sarcomatoid spindle cell carcinoma of the lungs (Figure 2).

The initial chest computerized tomographic scan was reported as stage IV advanced lung cancer (T4N2M0). The patient's clinical condition worsened within three months of diagnosis and four weeks waiting for the immunohistochemistry report and oncology review. She developed severe respiratory distress with Type 1 respiratory failure. A repeat chest X-Ray showed the total collapse of the left hemithorax. The clinical condition significantly deteriorated within 24 hours of re-admission, during which she succumbed to the illness while being prepared for admission into the intensive care unit (ICU).

Figure 2 A-D



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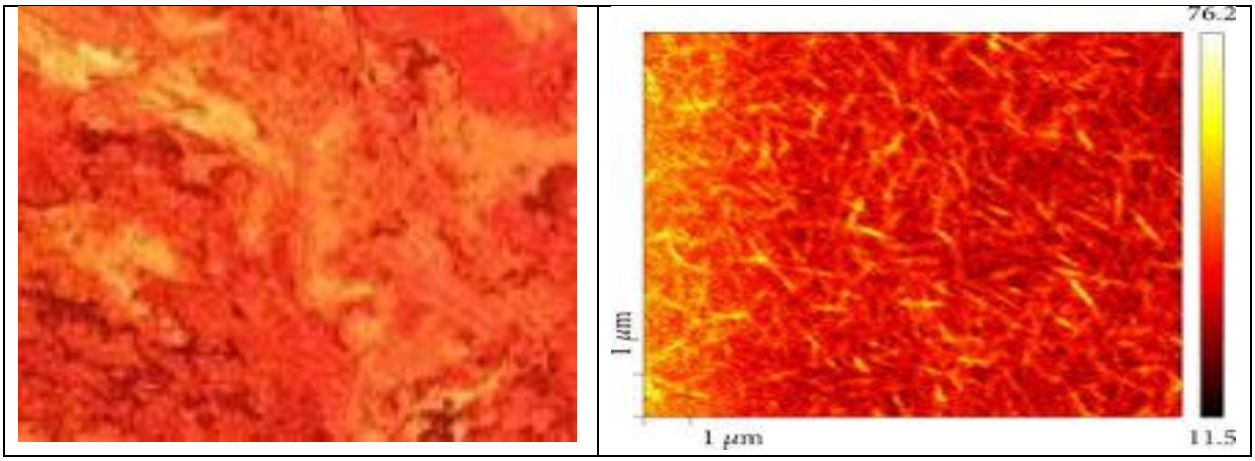

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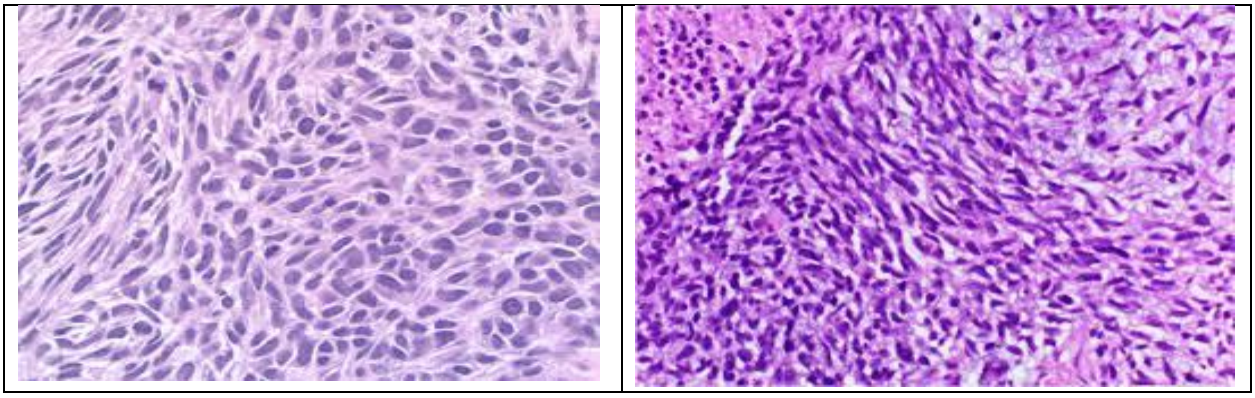

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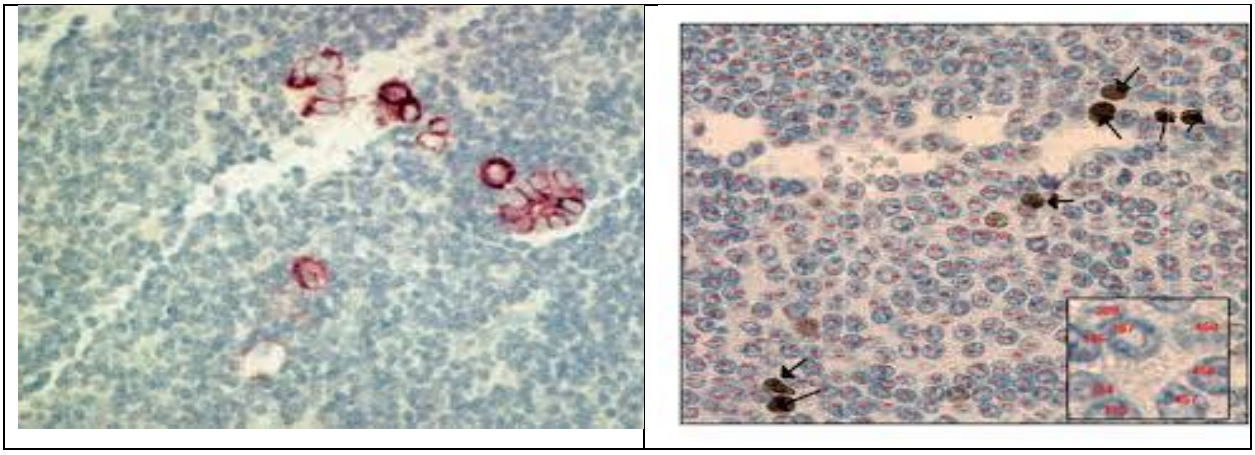

D

Histological analysis and immunohistochemical staining of lung biopsy.
(A) Haemotoxylin and eosin staining showed that the tumour cells are predominantly composed of spindle cells with increased mitotic activity and areas of necrosis (B) Tumor cells were weakly positive for P63 staining (C) Vimentin showed strongly positive tumour cells (D) Cytokeratin-Oscar showed tumour cells to be strongly positive.

Discussion

Sarcomatoid carcinoma of the lung is a subset of poorly differentiated non–small cell lung cancers, and the diagnosis can be challenging because they are uncommon. [1] These cancers predominantly affect male smokers, with a mean age at diagnosis of 65 years. [1] Sarcomatoid carcinomas are rare, comprising approximately 1% of all lung malignancies. [9] Typical symptoms include cough and haemoptysis, and the clinical course is usually aggressive as with conventional, non–small cell lung carcinoma. The stage of the disease is the most important prognostic indicator. [10] The index case occurred in a young, non-smoking female, which is atypical of the majority of the sarcomas of the lungs. The index patient had features that were well documented with the tumour.

The Chest computerized tomographic scan showed left-sided pleural effusion with pleural-based mass typical of the report in the literature. Radjev *et al*. reported a pedunculated mass on the right upper lobe of a 56-year-old woman with background Chronic Obstructive Pulmonary Disease (COPD), which showed spindle cell on histology. [11] Karmakar *et al.* also reported irregular margined mass in the left upper and middle lungs lobes, which showed spindle cells on histology in a 75-year-old retired teacher, an ex-smoker in India. [12] Sarcomatoid carcinomas may affect the central or peripheral part of the lungs but most commonly present as solitary, peripheral masses around the upper lobes, similar to other smoking-related, non–small cell carcinomas. [1]

The tissue biopsy with histology and immunohistochemistry assisted with making the diagnosis of the tumour of the index patient. The immunohistochemistry showed short, plum, spindle cells and broad-spectrum cytokeratins (AE1/3), typical of spindle cell sarcoma of the lungs. Radjev *et al.* demonstrated immunoreactivity for vimentin and Wilms Tumor-1 (WT-1), and focal positivity for pan-Cytokeratin (CK), AE1/AE3, P63, CK5/6, CK 8/18, CAM 5.2, Epithelial Membrane Antigen (EMA) and Gross Cystic Disease Fluid Protein-15 (GCDFP-15) for the diagnosis of a patient with spindle cell sarcoma. [11] However, Ouziane *et al.* reported tumour cells positivity for vimentin but negative for thyroid transcription factor-1 (TTF-1), epithelial membrane antigen (EMA), muscle actin (HHF35), cytokeratins 7, 20, and 5/6, actin, and calretinin in a 53-year-old woman. [13] On histology, the epithelial lineage of the spindle and giant cell components of pleomorphic carcinoma can often be demonstrated with multiple keratin antibodies, including AE1/3, CAM 5.2, CK18, and CK7, which are positive more frequently than epithelial membrane antigen, carcinoembryonic antigen (CEA), CD15, and Ber-EP4. [14] Keratin antibodies may highlight the epithelial component of carcinosarcoma but typically diffusely stain the epithelial component of pulmonary blastoma. [15] The positive ANA for the patient made the consideration for connective or inflammatory disease to be a close differential. However, tissue biopsy findings with histology and immunohistochemistry established the diagnosis of the tumour.

The definitive treatment could not be commenced in the index case because she died from respiratory failure complicating total left lung collapse while waiting for immunohistochemistry results and radio-oncology consultation. However, the treatment of spindle cell sarcoma of the lungs includes surgery and adjuvant chemotherapy. [16] Nodal involvement is a determinant prognostic variable because advanced stages are related to a worse prognosis because of early relapse of the disease. Surgery is recommended for N0 patients. [17] The aggressive nature of the disease in the index patient reduced the survival time to less than

three months from diagnosis. The prognosis of patients with pleomorphic carcinoma is poor, despite surgery and chemotherapy options. It has been observed that the survival rate at six months is only around 27%. The median survival time is about nine months to one year after a potentially curative surgical resection of the tumour. [11] The five-year survival rate is approximately 20% for sarcomatoid carcinoma compared with non-small cell lung cancer which has a five-year survival of 50%. [11]

Conclusion

Lung tumours are not all bronchogenic carcinoma. Although rare, spindle cell sarcomas may occur in very few lung cancer cases. Sarcomatoid carcinomas of the lung are a subset of poorly differentiated, non–small cell lung cancers. Clinically it is difficult to distinguish them from other tumours. The knowledge of the spindle cell sarcoma as one of the histologic types is important for considerations in the differential diagnosis of patients with lung tumours. The clinical history, imaging and pathologic findings are important in preventing misdiagnosis.

Authors' Contributions: OOT conceived the research drafted and revised the manuscript for sound intellectual content.
Conflicts of Interest: None.
Funding: Self-funded
Publication History: Submitted 26 December 2021**; Accepted** 16 March 2022**.**

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