International Journal of Science and Research (IJSR)
ISSN: 2319-7064
SJIF (2022): 7.942

Radiological Manifestations of Pulmonary Spindle Cell Lymphoma

Dr. Shweta Narwade[1], Dr. Dev Shetty[2], Dr. Ravi Verma[3]

Abstract: ***Spindle cell carcinoma is a rare and malignant variety of squamous cell carcinoma. It is a tumour that is constituted by a double cell proliferation: a carcinomatous sarcomatous epithelial cell of spindle cells and another sarcomatous spindle cell. It can affect any part of the body; however, it is more commonly found in the upper airway and digestive tract. It affects men more frequently between the sixth and seventh decade of life. It has aggressive behaviour with a tendency for recurrence. Alcohol and tobacco have been identified as the most important risk factors.***

Keywords: Radiologically, spindle cell lymphoma of the lung appears as a single large lesion, measuring 2 cm to 18 cm, middle lobe. Vascular invasion, spindle cell carcinoma

1. Introduction

Sarcomatoid neoplasms located in the lung and pleura are extremely rare, with an estimated incidence between 0.3 and 1.3% of all malignant lung neoplasms. They belong to a group of little differentiated non-small cell carcinoma that contain sarcoma, fusiform or giant cell components. According to the World Health Organization, it is classified into pleomorphic carcinoma, spindle cell carcinoma, giant cell carcinoma, carcinosarcoma and pulmonary blastoma. (1) The subtype of spindle cell carcinoma usually affects the oral cavity, larynx, breasts, kidney, uterus, conjunctiva, prostate, other organs, being rarely affected the lung. (2) Affected patients are predominantly males (4: 1) with a history of smoking, with an average age of approximately 65 years. The most common symptoms are cough and haemoptysis. The course of the disease is always aggressive, and the best indicator of prognosis is the stage. (3) The prognosis of patients with this entity is poor, with a 5-year survival rate of approximately 20%. (4)

Pulmonary spindle cell carcinoma is a type of non-small cell lung carcinoma. It is a variant of sarcomatoid carcinoma, which is a very rare case. (5) Spindle cell carcinoma has a prevalence of about 4.5–30.2% of all sarcomatoid carcinomas and represents only 0.4% of all primary lung malignancies. Demographically, the incidence is most commonly found in men compared to women (4: 1). The mean age at diagnosis of spindle cell carcinoma is ±62.5 years, with an age group of 30 years to 80 years. The risk factors are smoking and the effects of exposure to pollutants. (6) There is no standard treatment for this disease, though chemotherapy, surgery and radiation therapy may be considered. The prognosis is generally poor. The literature on disease progression and treatment are limited. (5) In this case report, we presented a diagnosed case of a 42-year-old female with pulmonary spindle cell lymphomabeing reported by histopathology.

Clinical details and imaging

This is a case of a 45 years old housewife, a resident of Mumbai who presented with a painless, progressively increasing lump in the left lower neck associated with grade-1 dyspnoea, occasional dry cough, loss of weight and loss of appetite since 6 months. A history of multiple joint pain was present. There was no history of fever, chest pain or any other comorbidities. The patient had no past history of pulmonary tuberculosis or multi drug resistant tuberculosis contact. A hysterectomy was done 10 years back in view of a prolapsed uterus. The vitals of the patient, systemic examination and routine blood investigations were all within normal limits except for tachycardia and grade 3 clubbing bilaterally. Chest radiograph was unremarkable. Neck ultrasound showed multiple necrotic supraclavicular lymphadenopathy and FNAC showed granulomatous necrotizing lymphadenopathy. The patient was started on an empirical anti-tubercular treatment (ATT) although the Genexpert was negative for MTB (Mycobacterium).

Around 2 months after starting ATT, the patient was hospitalized for increasing breathlessness. CECT thorax revealed a large heterogeneously enhancing mass with its epicentre in the pre-vascular space of the medias tinum and a few areas of necrosis within. It invaded the left innominate vein focally. Pericardial effusion was minimal. Discrete lymph nodes were observed in the left supraclavicular region, bilateral internal mammary regions, the right pre-vascular space, pre-caval region and the subcarinal region. Bilateral thin pleural effusions were seen. A PET/CT scan was then done followed by an appropriate CT-guided biopsy to differentiate between lymphoma, thymoma and germ-cell tumour, the most likely possibility benign lymphoma.

Ultrasound of the neck shows 3-4 necrotic lymph nodes (largest: 4cm x 1.8cm) in the left supraclavicular region and a TIRADS-2 lesion in the left hemi-thyroid. USG-guided FNAC of the cervical lymph nodes showed squamous and necrotic material while no growth was seen on the AFB smear and culture.
No abnormality was detected on ultrasound of the abdomen and pelvis (28/8/2022).

The TSH levels were within the normal range (2.29 mIU/L) while the other blood investigations such as d-dimer (0.69), RA factor (>8), CRP (165 mg/dL) and ESR (99mm/hr) were found to be raised.

The histopathological evaluation of the CT guided biopsy specimen revealed a spindle cell carcinoma (lymphoma).

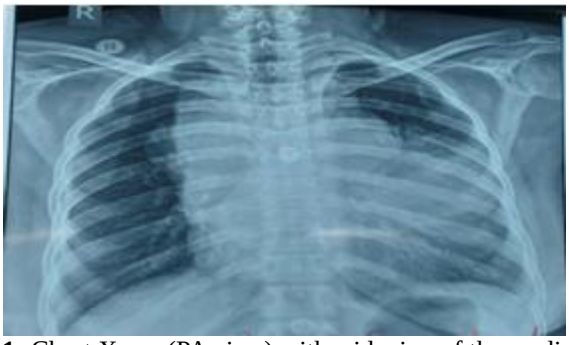

Figure 1: Chest X-ray (PA view) with widening of the mediastinum

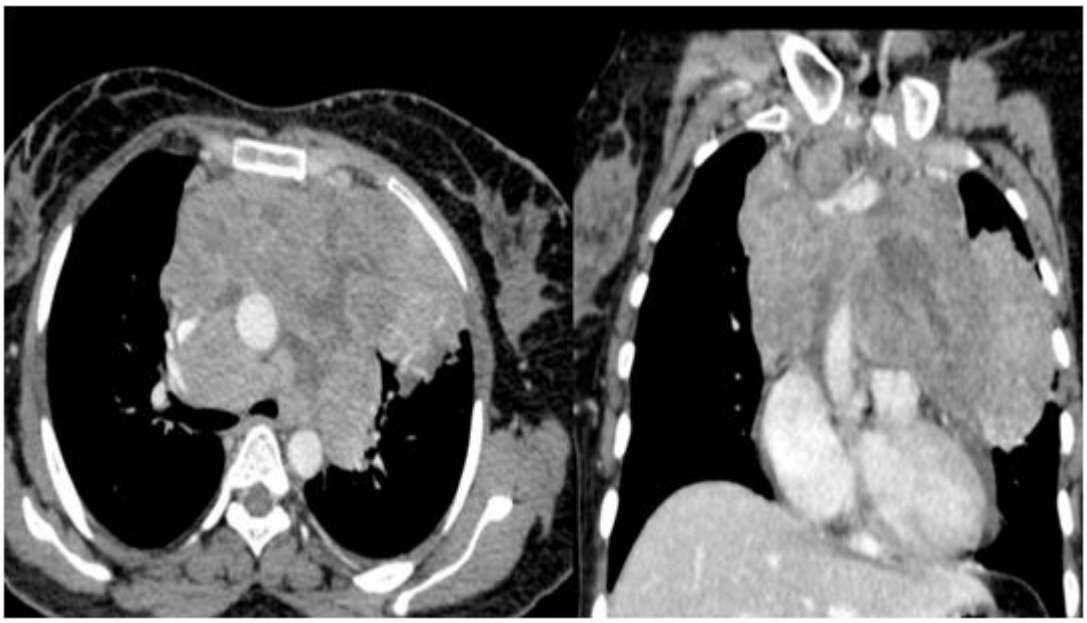

Figure 2: HRCT Thorax with a large heterogenous mass showing central necrosis

2. Discussion

Sarcomatoid carcinoma is the name given by WHO to unify pleomorphic carcinoma, spindle cell carcinoma, giant cell carcinoma, carcinosarcomas and blastomas. (4) These tumours are extremely rare with an estimated incidence of 0.3-1.3% of all lung malignancies. (7)

Pulmonary spindle cell carcinoma is a very rare case with nonspecific clinical symptoms and radiologic imaging similar to other lung cancers. Most cancer patients are asymptomatic in the initial stages and present only after stages 3 and 4 when they already have symptoms that interfere with the patient's activities. Shortness of breath, possibly persistent cough and sometimes accompanied by coughing up blood (haemoptysis), chest pain, voice changes, hoarseness, or loss of voice, changes in appetite leading to weight loss, individual fatigue, and headaches are a few symptoms. (8, 9)

This study reports a case of spindle cell lymphoma of a 42-year-old female patient with a history of dyspnoea, cough, loss of appetite and weight loss, which is related to what was written in the literature where the average age is 30 to 80 years although in a female which makes it a rarest condition. Symptoms depend on the location of the tumour. It has been described in cases of proximal tumours with cough, haemoptysis in 50% of cases, progressive dyspnoea and recurrent pneumonia due to bronchial obstruction more frequently, however, it can also be present. These patients present with chest pain in 25% of the cases of pleomorphic carcinoma. (10)

Radiologically, spindle cell lymphoma of the lung appears as a single large lesion, measuring 2 cm to 18 cm, often located peripherally in the middle lobe. Vascular invasion of spindle cell carcinoma is common in 40–70% of cases. (6) The imaging studies in our case are similar to reported literature in the form ofa large heterogeneously enhancing mass with its epicentre in the pre-vascular space of the mediastinum and a few areas of necrosis within and invasion of the left innominate vein focally. Pericardial effusion was minimal.

The preoperative diagnosis of spindle cell carcinoma is unknown in almost 60% of these cases due to the varying morphological numbers, therefore diagnosis is very difficult if only by sampling via FNAB. Surgery is an option to get a larger number of samples. Macroscopically, central necrosis and haemorrhage in spindle cell carcinoma of the lung are larger than 5 cm. (6)

Histologic characteristics of spindle cell carcinoma are homologous biphasic sarcomatoid carcinoma. Sarcomatoid areas in some biphasic spindle cell carcinoma contain numerous, cytologically bland, osteoclast-like giant cells. In

another variant histologic pattern, bluntly spindled tumour cells surround discrete zones of necrosis, imparting a necrotizing granuloma-like aspect to a portion of the proliferation. Immunohistochemistry markers are the parameter for a definitive diagnosis, that is highly positive for pan-cytokeratin, vimentin, and Ki67 with a mixture of monoclonal antikeratin antibodies (AE1/AE3, EMA EMA, CK). (9)

The treatment is similar to other NSCLCs, depending on the staging, and may include surgical resection, chemotherapy, and radiotherapy. Pulmonary spindle cell carcinoma response to pemetrexed and carboplatin. (5, 9) Spindle cell carcinoma is an aggressive tumour. Haemothorax in spindle cell carcinoma can occur due to the process of malignant angiogenesis, pulmonary infarction, necrotizing lung infection, hemopneumothorax, and coagulopathy disorders. The survival rate of spindle cell carcinoma patients is about five years, slightly shorter than other types of NSCLC. (11)

3. Conclusion

A 42-year-old female presented with dyspnoea, cough, weight loss and loss of appetite without any comorbidities. The symptoms worsened in spite of starting empirical ATT. Hospitalization was required for further management. On Radiological investigations the final diagnosis of the patient was lympjoma, therefore radiological investigations are of vital importance for a timely diagnosis to begin surgical management accompanied by chemotherapy and radiotherapy.

In tuberculosis endemic countries, it is not uncommon for necrotic lymphadenopathy to be treated with empirical anti tuberculous therapy. Lack of clinical response should prompt a high index of suspicion to search for an underlying neoplasm. Radiological investigations, image guided biopsy, histopathology and immunohistochemistry play a critical role in the management of such cases.

References

[1] Travis WD. Sarcomatoid neoplasms of the lung and pleura. Arch Pathology Lab Med [Internet].2010 Nov [cited 2023 Apr 14]; 134 (11): 1645–58. Available from: https: //pubmed. ncbi. nlm. nih. gov/21043818/

[2] Qi DJ, Liu B, Feng L, Zhao L, Yan P, Du J, et al. Pulmonary spindle cell carcinoma with unusual morphology: A rare case report and review of the literature. Medicine [Internet].2017 Jun 1 [cited 2023 Apr 14]; 96 (24). Available from: /PMC/articles/PMC5478320/

[3] Franks TJ, Galvin JR. Sarcomatoid carcinoma of the lung: histologic criteria and common lesions in the differential diagnosis. Arch Pathology Lab Med [Internet].2010 Jan 1 [cited 2023 Apr 14]; 134 (1): 49–54. Available from: https: //pubmed. ncbi. nlm. nih. gov/20073605/

[4] Ávila Martínez RJ, Marrón Fernández C, Hermoso Alarza F, Zuluaga Bedoya M, Meneses Pardo JC, Gámez García AP. Carcinomas Sarcomatoides Pulmonares Primarios. Arch Bronco pneumology.2013 Sep 1; 49 (9): 405–7.

[5] Sardar MA, Azharuddin M, Khan WJ, Noory MA, Shaikh N, Ullah Malik S, et al. USA 5. Department of Internal Medicine. Monmouth Medical Center.

[6] Tsuta K, Kalhor N, Wistuba II, Moran CA. Clinicopathological and immunohistochemical analysis of spindle-cell carcinoid tumour of the lung. Histopathology [Internet].2011 Sep 1 [cited 2023 Apr 15]; 59 (3): 526–36. Available from: https: //onlinelibrary. wiley. com/doi/full/10.1111/j.1365-2559.2011.03966. x

[7] Terada T. Spindle cell carcinoma of the lung: Frequency, clinical features, and immunohistochemical studies of three cases. Respir Med CME.2010 Jan 1; 3 (4): 241–5.

[8] Kontic M, Stojsic J, Stevic R, Bunjevacki V, Jekić B, Dobricic V. Could spindle cell lung carcinoma be considered and treated as sarcoma, according to its clinical course, morphology, immunophenotype and genetic finding? Pathology and Oncology Research [Internet].2013 Aug 26 [cited 2023 Apr 15]; 19 (1): 129–33. Available from: https: //link. springer. com/article/10.1007/s12253-012-9562-4

[9] Feng L, Cai D, Muhetaer A, Yang YL, Ren F, Yishake M, et al. Spindle cell carcinoma: the general demographics, basic clinico-pathologic characteristics, treatment, outcome and prognostic factors. Oncotarget [Internet].2017 May 19 [cited 2023 Apr 15]; 8 (26): 43228–36. Available from: https: //www.oncotarget. com/article/18017/text

[10] Feki W, Ketata W, Yangui I, Kammoun S. Spindle Cell Carcinoma of the Lung: Benefits of Immunohistochemical Studies. J Pulm Respir Med.2018; 8 (1): 449.

[11] Shoji F, Maruyama R, Okamoto T, Ikeda J, Nakamura T, Wataya H, et al. Long-term survival after an aggressive surgical resection and chemotherapy for stage IV pulmonary giant cell carcinoma. World J Surg Oncol [Internet].2005 Jun 2 [cited 2023 Apr 15]; 3 (1): 1–4. Available from: https: //wjso. biomedcentral. com/articles/10.1186/1477-7819-3-32