Primary Pulmonary Spindle Cell Carcinoma in a Young Nonsmoker Patient

Abstract

Spindle cell carcinoma (SCC) is a very rare type of tumor. Very few case reports of primary pulmonary SCC have been reported in the literature. It is a type of sarcomatoid carcinoma with very poor prognosis. It is usually seen in patients in the age group of 50–80 years. Exact etiology is unknown till now. We report a rare case of primary pulmonary SCC in a young nonsmoker patient who presented in a very advanced stage.

Keywords: *Lung cancer, pulmonary, spindle cell carcinoma*

Amitesh Gupta, Parul Mrigpuri

Department of Pulmonary Medicine, Shri Lal Bahadur Shastri Government Medical College, Ner Chowk, Himachal Pradesh, India

Introduction

Spindle cell carcinoma (SCC) is a rare type of lung cancer representing 0.2%–0.3% of all primary pulmonary malignancies. There is male-to-female predominance (4–5:1), common in smokers, and in patients between 50 and 80 years of age.[1] It does not contain any differentiated carcinomatous component but consists only of pure "malignant" spindle cells. Pulmonary SCC is classified as sarcomatoid carcinoma of the lung, which is an aggressive tumor, the other subtypes being pleomorphic carcinoma, giant cell carcinoma, carcinosarcoma, and pulmonary blastoma.[2] Sarcomatoid tumors of the lung together comprise <1% (between 0.1% and 0.4%) of all reported lung cancer cases. SCC of the lung is a very rare type that can develop anywhere in the lung. The cause of cancer is generally unknown, but smoking is found to be one major risk factor. The presenting symptoms are chest pain, shortness of breath, fatigue, and other general signs and symptoms, such as fever, weight loss, and appetite loss. Chemotherapy, surgery, radiation therapy, and other palliative measures may be used for treating SCC of the lung depending on the staging. The prognosis may depend on many factors including the stage of the tumor, progression, response to treatment, and overall health of the individual. However, in general, the prognosis of SCC of lung is poor.[3] We report a rare case of primary pulmonary SCC in a young nonsmoker patient.

Case Report

A 28-year-old male nonsmoker, daily laborer, came with complaints of left-sided chest pain for 6 months, shortness of breath and cough for 3 months. General physical examination was normal. Laboratory investigations showed total leukocyte count to be 13,000/dl with normal liver and renal function tests. On systemic examination, bulge was seen on inspection on the left side of chest with absent breath sounds on auscultation. Chest X-ray showed an opaque left hemithorax with shift of mediastinum to opposite side [Figure 1]. The patient was already on the first-line antitubercular treatment (ATT) started on the basis of chest X-ray, but there was no improvement in symptoms. There was no history of drug abuse or smoking in the past. Medical history was unremarkable except for this episode. There was no history of malignancy in family.

Pleural tapping showed a hemorrhagic fluid with predominant lymphocytes but low adenosine deaminase levels. Contrast-enhanced computed tomography (CECT) of the chest was done which showed a mass of heterogenous density with solid and cystic components occupying whole of left hemithorax and herniating to the right after crossing the midline [Figure 2]. The mass was also displacing the heart and great vessels with involvement of left pericardium. Anteriorly, it was eroding the ribs and reaching chest wall, thus depicting its malignant nature. Bronchoscopy showed left upper and lower lobe bronchus to be completely occluded

How to cite this article: Gupta A, Mrigpuri P. Primary pulmonary spindle cell carcinoma in a young nonsmoker patient. Clin Cancer Investig J 2018;7:116-8.

Address for correspondence:
Dr. Parul Mrigpuri, Department of Pulmonary Medicine, Shri Lal Bahadur Shastri Government Medical College, Mandi, Ner Chowk - 175 021, Himachal Pradesh, India.
E-mail: parul57_mrigpuri@yahoo.com

Access this article online

Website: www.ccij-online.org

DOI: 10.4103/ccij.ccij_5_18

Quick Response Code:

due to external compression with no endobronchial growth. Fine needle aspiration cytology was done which showed hemorrhagic smears.

Biopsy was taken from the lung mass, and the sections revealed spindle-shaped cells arranged in intersecting fascicles with focal areas of necrosis and high mitotic activity consistent with a malignant spindle cell tumor [Figure 3].

Due to highly advanced stage and involvement of major vital structures, surgery was not an option and the patient was given option for referral to advanced oncology center, but the patient's relatives refused for further treatment.

Discussion

Sarcomatoid tumors of the lung together comprise <1% (between 0.1% and 0.4%) of all reported lung cancer cases. SCC of the lung is a very rare type of lung cancer. Individuals below the age of 40 years are rarely diagnosed with SCC. To the present knowledge of the author, no case has been reported so far in a patient below 30 years of age as was our patient.[1]

Our case was a young, nonsmoker patient who was not improving on ATT. A high index of suspicion was kept, and the patient was investigated further.

The exact cause of SCC of the lung is unknown. However, it is believed that there are certain factors such as smoking, exposure to radon, and prolonged exposure to asbestos and other harmful chemicals (arsenic, chromium, nickel, and tar), chromosomal anomalies, and mutations on gene TP53 that may contribute to SCC risk factors.[4] Signs and symptoms seen in early stages may lead to misdiagnosis for infective conditions. In later stages, they may invade pleura, chest wall, and mediastinum and disseminate through blood and lymphatic system to distant sites. Badshah *et al*. reported one case of spindle cell sarcoma which presented with Pancoast syndrome.[5] Diagnosis can be made by keeping high index of suspicion in a patient not responding to conventional treatment particularly if treatment has been started empirically. Diagnostic modalities such as chest X-ray, CECT, magnetic resonance imaging, and positron emission tomography may be used to reach the diagnosis although confirmation can be made only after histopathology. Immunohistochemical features of SCC are unknown; however, in one series of three cases reported by Terada, pan-cytokeratin, CK18, and vimentin were positive in all three.[6] Treatment options available for individuals with SCC of the lung are dependent upon site and extent of involvement. In Stage 1, SCC presents as a small lump which can be excised and carries the best prognosis. In Stage 2 and Stage 3, micro-metastasis occurs and has poor prognosis. The most commonly used treatment is surgery. Surgery can be potentially curative if the tumor is completely excised (in case of early-stage tumors). However, some cases show recurrence many years later.

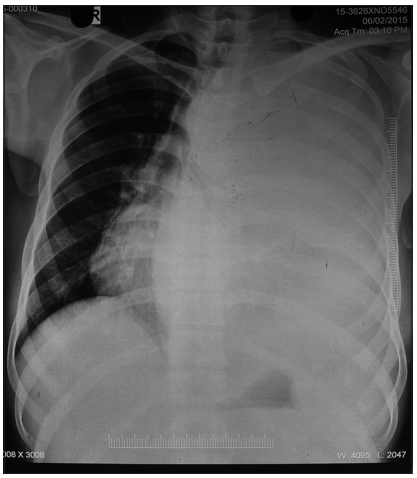

Figure 1: Chest X-ray showing left opaque hemithorax with mediastinal shift to the right

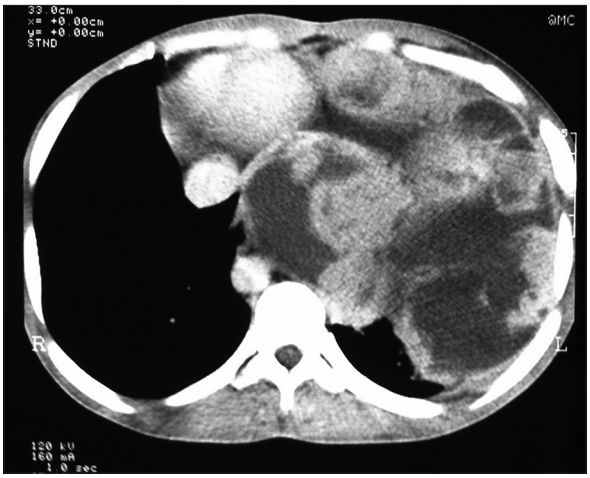

Figure 2: Contrast-enhanced computed tomography chest showing heterogenous mass in the left hemithorax

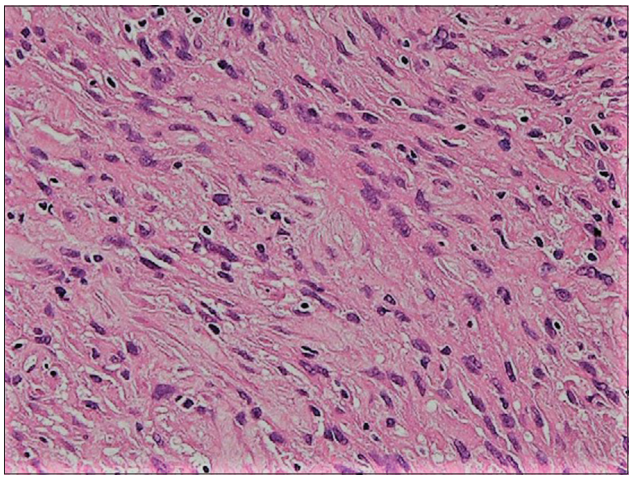

Figure 3: Spindle cell carcinoma histopathology (×100)

Chemotherapy and radiation may also be used for treatment, if surgery is not a viable option, or if there is a suspicion of metastasis. SCC is a type of aggressive malignancy. The

prognosis of the condition is generally poor. The death rate is high even when cancer is diagnosed at an early stage.[7]

Conclusion

SCC is a rare form of lung cancer representing 0.2%–0.3% of all primary pulmonary malignancies. These tumors are associated with a poor prognosis even with combined surgery, chemotherapy, and radiation therapy, and only 10% of patients survive 2 years after diagnosis.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

References

1. Dail DH, Hammar SP. Pulmonary Pathology. 2nd ed. New York: Springer-Verlag; 1994. p. 1310-2.
2. Travis WD, Brambilla E, Burke AP, Marx A, Nicholson AG, editors. WHO Classification of Tumours of Lung, Pleura, Thymus and Heart. 4th ed. Lyon: IARC; 2015.
3. Nappi O, Glasner SD, Swanson PE, Wick MR. Biphasic and monophasic sarcomatoid carcinomas of the lung. A reappraisal of 'carcinosarcomas' and 'spindle-cell carcinomas'. Am J Clin Pathol 1994;102:331-40.
4. Weissferdt A, Kalhor N, Rodriguez Canales J, Fujimoto J, Wistuba II, Moran CA, *et al.* Spindle cell and pleomorphic ("sarcomatoid") carcinomas of the lung: An immunohistochemical analysis of 86 cases. Hum Pathol 2017;59:1-9.
5. Badshah A, Khan S, Saeed U. Spindle cell sarcoma presenting as Pancoast syndrome. J Coll Physicians Surg Pak 2016;26:623-5.
6. Terada T. Spindle cell carcinoma of the lung: Frequency, clinical features, and immunohistochemical studies of three cases. Respir Med CME 2010;3:241-5.
7. Rossi G, Cavazza A, Sturm N, Migaldi M, Facciolongo N, Longo L, *et al.* Pulmonary carcinomas with pleomorphic, sarcomatoid, or sarcomatous elements: A clinicopathologic and immunohistochemical study of 75 cases. Am J Surg Pathol 2003;27:311-24.