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Case Report

A case report of primary pleomorphic lung cancer with an atypical presentation

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ABSTRACT

Pulmonary pleomorphic carcinoma (PPC) is an uncommon primary lung tumor with a low incidence and aggressive biological characteristics that can occur in young people and nonsmokers. Pleomorphic carcinoma (PC) has a worse prognosis than other non-small cell lung tumors. We describe a 45-year-old man who complained of having a dry cough for a month. A well-defined mass in the anterior section of the left lower lobe was identified by chest high-resolution computed tomography (HRCT), and a positron emission tomography (PET) scan confirmed a left lobulated tumor with fluorodeoxyglucose (FDG) uptake in the left mediastinal lymph nodes. An endobronchial ultrasound guided transbronchial needle aspiration (EBUS TBNA) guided biopsy of the tumor was performed and sent for histopathological analysis, which identified pleomorphic squamous cells and giant cells. Positive IHC markers included P63, TTF1, pancytokeratin, Vimentin. Patient was diagnosed with stage IIIa (T3N2M0) pleomorphic carcinoma of the lung. Due to the tumor's advanced stage, the patient received chemotherapy and radiation therapy.

Keywords: IHC markers, PET scan, Pleomorphic pulmonary carcinoma, Rare primary lung carcinoma

INTRODUCTION

Pleomorphic carcinoma (PC), a rare malignant lung tumor, is estimated to account for 0.1% to 0.4% of all lung cancers.[1] The WHO classified PC as a subtype of lung cancer with pleomorphic, sarcomatoid, or sarcomatous components in 2004.

PC is classified as a poorly differentiated adenocarcinoma, squamous cell carcinoma, large cell carcinoma with sarcomatoid components of the spindle or giant cells in a minimum of 10% of the cells, or a carcinoma made entirely of giant and spindle cells.[2,3] According to the WHO classification of 2021, PC is a subtype of sarcomatoid carcinoma, whereas PC is subdivided into three subtypes: pleomorphic carcinoma, giant cell carcinoma, and spindle cell carcinoma.[4]

PC has a more severe clinical course and a worse prognosis when compared to other non-small cell lung cancers.[5,6] PC might be found anywhere in the body, although the respiratory system is accountable for the bulk of cases. The tumor that grows from the respiratory system is known as pulmonary pleomorphic carcinoma.

The average patient age at diagnosis is between 60 and 65 years old, and 60% to 90% of patients had a history of smoking, indicating a strong connection with smoking.[5-7] In this report, we describe a pulmonary pleomorphic carcinoma (PPC) case involving a young male nonsmoker who presented to the OPD with a dry cough and was later confirmed with the disease.

CASE REPORT

A 45-year-old lecturer visited the pulmonology outpatient department after complaining of a dry cough for a month. He did not report any symptoms of hemoptysis, shortness of breath, chest pain, or lack of appetite. He does not smoke and has no medical conditions. There is no remarkable family history. He has no previous history of tuberculosis, bronchial asthma, or chronic respiratory illness. During the examination, vital signs were steady, and auscultation verified typical vesicular breath sounds with no additional noises.

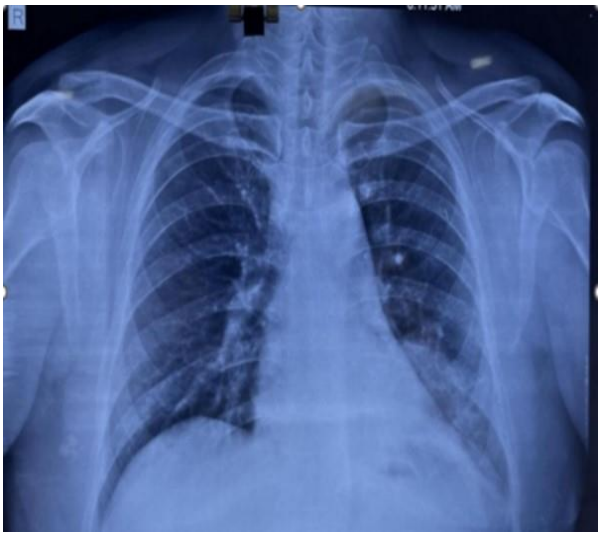

Figure 1: Chest X-ray showing left mid zone opacity.

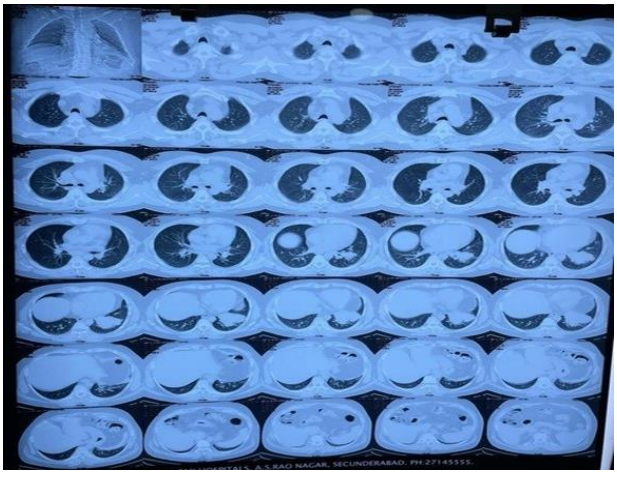

Figure 2: HRCT chest showing large lobulated dense opacity in the left lower lobe.

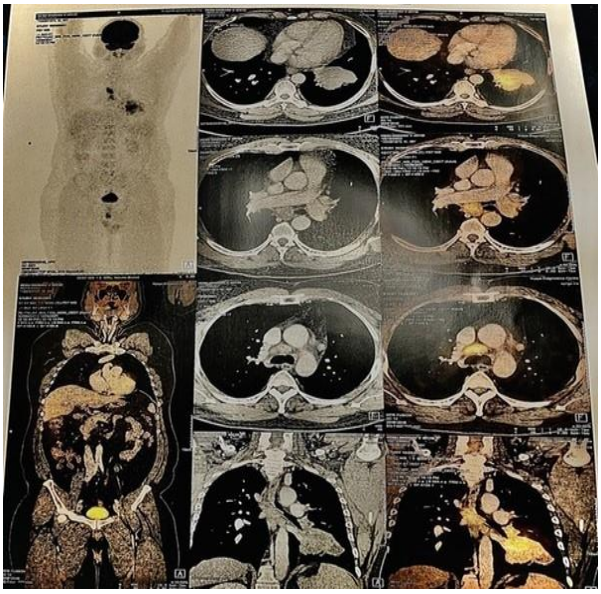

Figure 3: PET scan reveals increased FDG uptake in left lower lobe, increased FDG uptake in left hilar, subcarinal, and pre/paratracheal lymph nodes.

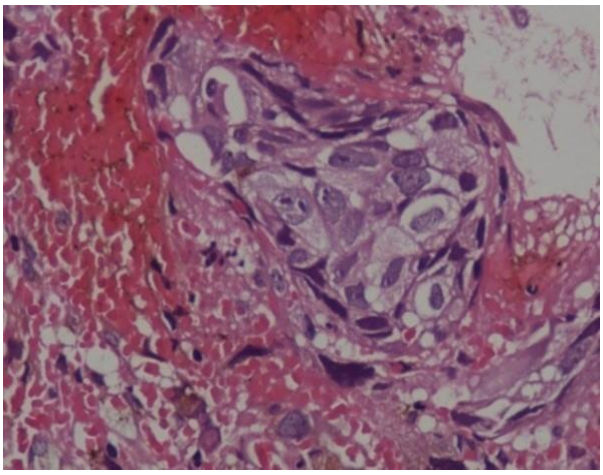

Figure 4: Pleomorphic cells.

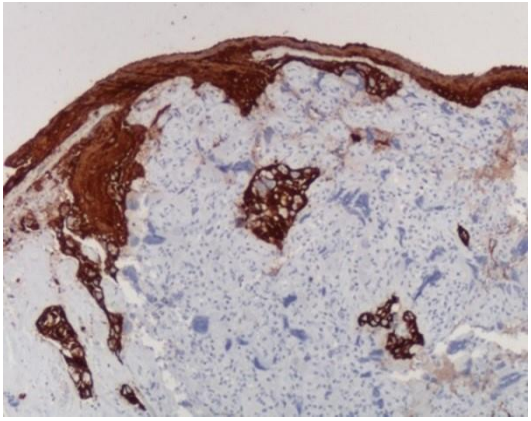

Figure 5: Pan cytokeratin positive pleomorphic carcinoma.

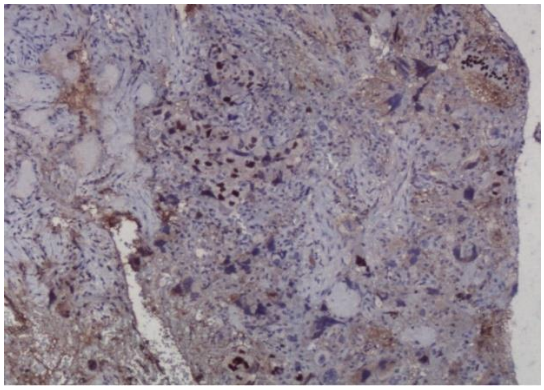

Figure 6: P63 positive.

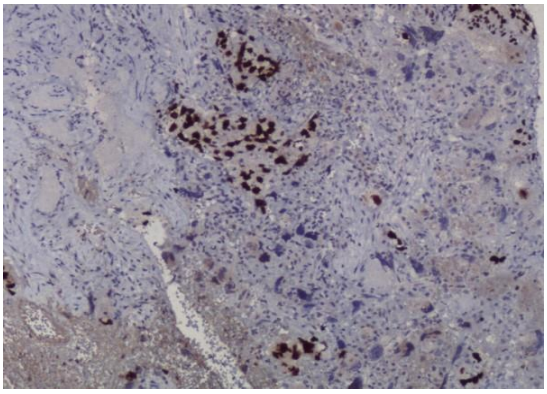

Figure 7: TTF1 positive.

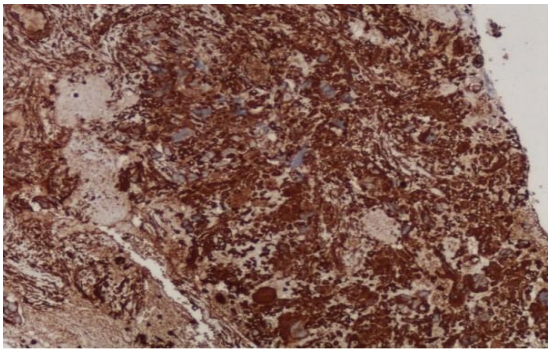

Figure 8: Vimentin positive.

A chest X-ray indicated opacity in the left midzone (Figure 1). High-resolution computed tomography (HRCT) detected a well-defined soft tissue density lesion of (76×59) mm in the anterior basal region of the left lower lobe (Figure 2).

A PET scan indicated a lobulated mass measuring 6.9×4.33×4.7 cm in the lower lobe of the left lung, as well as fluorodeoxyglucose (FDG) uptake in the left hilar, subcarinal, pericranial, and paratracheal lymph nodes (Figure 3).

Endobronchial biopsy was done on the mass, and histology was performed on endobronchial ultrasound guided transbronchial needle aspiration (EBUS TBNA) guided mediastinal lymph nodes, which indicated pleomorphic squamous cells and giant pleomorphic cells (Figure 4). Pan cytokeratin (Figure 5), P63 (Figure 6), TTF1 (Figure 7), and Vimentin (Figure 8) were found to be positive, but desmin was found to be negative.

Over two weeks, he was thoroughly examined and diagnosed with stage IIIa (T3N2M0) pleomorphic carcinoma of the lung.

For six weeks, the patient received radiation therapy once a week at a dose of 6000 Em/30 fr. Chemotherapy (injection cisplatin and injection etoposide) was administered once a day for six weeks. A six-week follow-up PET scan revealed no change in the size of the lung tumor, as well as FDG uptake in the brain and an increase in FDG uptake in the lymph nodes, indicating a bad prognosis due to the aggressive nature of the tumor.

DISCUSSION

PC is a rare and aggressive type of lung cancer. Previous research has found that old people, with an average age of 60 to 65 years, are more likely to get PPC, which has a high male prevalence and is connected to smoking.[5-7] There have been reports of PPC patients from many ethnic groups. The specific incidence in different races was not published due to the small number. A recent study, however, found that nearly 80% of PPC patients were white.[8] PPC patients have reported symptoms such as chest pain, irritating cough, hemoptysis, fever, and weight loss.[9]

The patient in the study, on the other hand, is a young male non-smoker, which is not a typical presentation for the tumor. It is crucial to point out that the incidence of pleomorphic cancer in a young, nonsmoking man with a different presentation cannot be ruled out. Even if examinations indicate that the tumor is aggressive in its advanced stages, patients may be asymptomatic or present with mild symptoms, such as a 1-month dry cough in this patient. Presentations like these, with minimal symptoms, are typically unrecognized until the later stages of the tumor, so a thorough clinical history, physical examination, and, if necessary, investigations are crucial for early detection and treatment. Due to the limits of biopsy samples, pleomorphic carcinoma can be difficult to diagnose before surgery. Spindle cell carcinoma, giant cell carcinoma, carcinosarcoma, and pulmonary blastoma are the other four types of sarcomatoid cancer. Most lesions presented as peripheral masses with significant enhancement on contrast CT images. Lesions larger than 5 cm in diameter commonly had myxoid degeneration and central low attenuation zones, indicating hemorrhage or necrosis. Cavitary lesions were frequently found as a result of central necrosis.[10]

Lung pleomorphic carcinomas can spread to other organs such as the brain, liver, adrenal tissue, and bone. PPC has a more malignant biological behavior than other non-small cell lung cancer subtypes; even early tumors may penetrate

blood vessels, and early pulmonary pleomorphic carcinoma with no lymphatic dissemination can return or spread. Several previous studies involving a large number of patients discovered that TNM staging is an important predictive tool. Still, there is no standard strategy for treating pleomorphic cancer. While surgery had no influence on survival in patients with advanced tumors, it significantly improved survival in those with localized tumors. So, for those with early-stage pulmonary pleomorphic carcinoma, surgery is the treatment of choice.[11,12]

There have been a few studies on chemotherapy for the early stages of PPC in the literature, and multiple randomized clinical trials have shown that the efficiency of adjuvant chemotherapy in treating early non-small cell lung cancer (NSCLC) positron emission tomography (PET) scan is also ambiguous.[13,14] PPC patients who received chemotherapy had a worse prognosis than those who did not in the localized stage, which could be due to the cytotoxicity of chemotherapy medicines. As a result, chemotherapy remains a controversial treatment option for early pulmonary pleomorphic cancer. According to one study, the overall median survival in postoperative, recurring, and inoperable patients who received cytotoxic treatment was around 5 months.[15]

Cisplatin and vinorelbine may be effective as neo-adjuvant treatments for PPC patients.[16] The therapeutic effects of radiation therapy for PPC have not been well researched. Patients with epidermal growth factor receptor (EGFR) mutations may benefit from personalized therapy.[17] In recent years, novel targeted treatments and PD-1 inhibitors have introduced new therapeutic options for advanced stages of carcinoma.[18] As our patient's tumor was in an advanced stage and he was not a suitable candidate for surgery, he got 6 weeks of radiotherapy, cisplatin, and etoposide chemotherapy.

CONCLUSION

The most common type of pleomorphic carcinoma is pulmonary pleomorphic carcinoma. PC is still associated with a worse prognosis than other non-small cell lung tumors. Non-smokers might also develop pulmonary pleomorphic cancer. A case of pleomorphic pulmonary carcinoma in a non-smoker male patient under 60 has been reported. Doctors face a significant challenge in recognizing unusual appearances, which can have catastrophic consequences for patients' health because many cases are detected in the later stages when the patient's treatment options are restricted. Patients must be thoroughly examined and followed up on to find such less common tumors early and avoid them from going unnoticed.

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