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Case Report

Case Report of Adult Pulmonary Blastoma: Very rare presentation

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ABSTRACT

A case study of a 41-year-old male patient, a known smoker presented with complaints of right-sided chest pain, cough and shortness of breath for two months for which he got a chest X-ray, which revealed soft tissue opacity in the right upper and middle lobe. On examination there were slightly diminished breath sounds over the right hemi thorax. A pulmonary blastoma (PB) diagnosis was confirmed through a biopsy and immunohistochemistry. Diagnostic imaging (CT, MRI, PET-CT) was done as it plays a crucial part in seeing the tumor resectability, treatment response and rule out metastatic disease. Although surgical excision is the treatment of choice in PB but in our case, the tumor was extremely aggressive, progressive and irresectable so chemotherapy was recommended by a multidisciplinary specialized team.

Keywords: Pulmonary blastoma, Pleuropulmonary blastoma, Fetal adenocarcinoma, Immunohistochemistry and biopsy, resectability, Chemotherapy.

How to Cite This Article: *Asghar A, Rauf MH, Sheikh AZ, Siddique K, Hassan A. Case Report of Adult Pulmonary Blastoma: Very rare presentation. Pak Armed Forces Med J 2026; 76(Suppl-7): S1200-S1201. DOI: https://doi.org/10.51253/pafmj.v76iSUPPL-7.12592*

INTRODUCTION

Pulmonary blastoma is exceedingly rare lung cancer, which is only 0.25-0.5% of all lung cancer cases.[1,2] It is segregated into three subtypes: classic biphasic PB (CBPB), well-differentiated fetal adenocarcinoma (WDFA), and pleuropulmonary blastoma. CBPB, the most common subtype, is characterized by mix epithelial and mesenchymal cancer cells[3]. Unfortunately, the prognosis is poor, with a 5-year survival rate of around 15-16%[4]. We present a unique case of pulmonary blastoma with progression in size and is currently inoperable. Chemotherapy was recommended, with a plan to reassess the patient's status after treatment.

CASE REPORT

41-year-old male patient, presented with complaints of pain on right side of the chest and shortness of breath; however, there were no complaints of weight loss, anorexia or hemoptysis. X-ray showed soft tissue mass in the right upper and middle lobe. CT-guided biopsy from outside the hospital was reported as non-small cell lung carcinoma. The block submitted for review in our institute showed scanty focus of adenocarcinoma. Prognostic markers as well as molecular testing could not be performed due to its scant nature. CT-guided biopsy was again performed and evaluated at the Shaukat Khanam Memorial Cancer Hospital & Research Centre (SKMCH&RC) in Lahore Pakistan (Figure-1).

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Received: 21 Aug 2024; revision received: 15 Oct 2024; accepted: 16 Oct 2024

IMMUNO/HISTOCHEMICAL STAIN(S):

INSM1:	Negative
Napsin:	Focal positive
Ki67:	5-10% proliferation index
TTF1:	Focal positive
Synaptophysin:	Negative
Desmin:	Negative
SS18-SSX:	Negative
B catenin:	Focal positive (nuclear staining)

Figure-1: Immunohistochemistry Report of the Patient

PET CT scan of the whole body revealed a fluorodeoxyglucose (FDG) avid lobulated large heterogeneously enhancing lung mass in right upper and middle lobes, measuring 13.4 x 7.6 x 11.9 cm, SUV 12.9. It was invading the chest wall anteriorly as shown in Figure 2 (A, B & C). A chest radiograph after one month showed soft tissue density mass in the right middle zone. Follow-up chest radiograph after 15 days shows disease progression as shown in Figure-2 (D).

CE-CT chest and abdomen demonstrate interval increase size measuring 19 x 11 x 27 cm, extending across the midline with a mediastinal shift and displacing the right hemi diaphragm and liver inferiorly Figure-2 (E). The involvement of mediastinum makes it T4N0M0 disease. CT scan after 3 cycles of chemotherapy (Carboplatin + Paclitaxel x

Cycle 3) shows a marginal decrease in size and after completion of 9 cycles, it demonstrates further interval decrease in size as shown in Figure-2 (F).

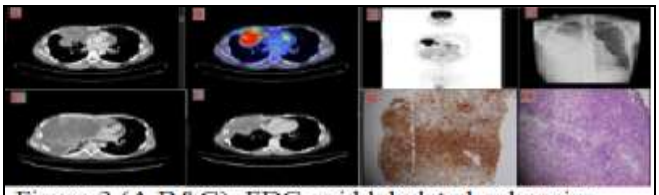

Figure-2 (A,B&C). FDG avid lobulated enhancing lung mass in right upper and middle lobes (D) X ray chest shows progression of the disease.(E) Mass is extending across the midline with mediastinal shift, displacing the right hemi diaphragm and liver inferiorly.(F) post treatment scan after 9 cycles of chemotherapy, interval decrease size of mass.(G&H) Histopathology slides from the biopsy of lesion showing malignant neoplasm with a biphasic morphology, comprised in a hyper cellular spindled, acinar pattern with mesenchymal component

DISCUSSION

Pulmonary blastoma is declared as a separate subtype of sarcomatoid carcinoma. Almost 80% of patients diagnosed with CBPB and WDFA are normally have a smoking history, and most of them are in the age group of forty and fifties[5].In case of localized disease surgical resection is the prime choice of treatment.[6,7] Due to rarity of CBPB, chemotherapy regimen is undefined. There is requirement of multidisciplinary collaborative treatment in order to improve survival rates because of poor prognosis in CBPB8. It is recommended that including a platinum based agent and Etoposide (VP-16) can yield a relatively satisfactory response[2]. Our patient had received 9 cycles of chemotherapy (Carboplatin + Paclitaxel) that were well tolerated. For the management of unresectable tumor, the efficacy of radiotherapy has been established for locally aggressive and metastatic disease. There is a high rate of recurrence of about 43% during the first year after treatment and with inclination for metastasizing to the mediastinum, pleura, diaphragm, liver, heart, soft tissues and brain[2].

To summarize, the intricate histological features of classic biphasic pulmonary blastoma (CBPB) pose a significant diagnostic challenge, particularly when working with limited biopsy samples, emphasizing the need for precise and reliable diagnostic techniques. Timely treatment is essential for classic biphasic pulmonary blastoma (CBPB), to improve survival chances and prevent severe complications like intratumoral bleeding, as observed in our patient.

Conflict of Interest: None.

Funding Source: None.

Authors' Contribution

Following authors have made substantial contributions to the manuscript as under:

AA & MHR: Data acquisition, data analysis, critical review, approval of the final version to be published.

AZS & KS: Study design, data interpretation, drafting the manuscript, critical review, approval of the final version to be published.

AH: Conception, data acquisition, drafting the manuscript, approval of the final version to be published.

Authors agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved.

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