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Case Report

CASE REPORT

# Metachronous pleuropulmonary blastoma in an adult patient with endometrial cancer: a case report

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## Abstract

Pulmonary blastoma is a rare form of lung cancer with a reported incidence of 0.25–0.55 of primary pulmonary cancers. Pleuropulmonary blastoma (PPB) is a common finding in children while it is rarely found in adults. In the past few years, the incidence of a second primary tumour has increased to 3.5% followed by third primary tumour at 0.5% and fourth tumour at 0.3%. The clinical significance of diagnosing and distinguishing a secondary primary tumour is often challenging. As per our knowledge, this is the first case of metachronous PPB in an adult patient previously diagnosed with endometrial cancer.

## INTRODUCTION

Pulmonary blastoma is an uncommon form of lung cancer. As per current evidence, it accounts for 0.25–0.5% of primary pulmonary cancers [1]. Pulmonary blastoma is classified into three subtypes, i.e. biphasic pulmonary blastoma, well-differentiated foetal adenocarcinoma and malignant pleuropulmonary blastoma (PPB) [1]. Due to its rarity, the interpretation of its published clinical features and epidemiology is perplexing. We present an unusual case of PPB in an adult patient with endometrial cancer.

## CASE REPORT

A 56-year-old patient presents to the hospital with complaints of general weakness and shortness of breath. The patient was diagnosed with a grade II well-differentiated adenocarcinoma of the endometrium in February 2014. Considering the patient's history of adenocarcinoma of the endometrium, a whole-body positron emission tomography/computed tomography (PET-CT) was carried out in August 2018 to assess disease progression or metastases.

The PET-CT scan revealed a hypermetabolic heterogeneously enhancing soft-tissue mass in the right upper lobe (Fig. 1). Well-defined heterogeneously enhancing soft-tissue lesion measuring 85 × 84 × 83 mm was FDG avid with an SUV max 22.3 in the right upper lobe. It was causing cut off of the apical segmental bronchus,laterally abutting the pleura and medially invading the mediastinal pleura. The lesion shows few areas of central and peripheral necrosis. There is no obvious associated rib destruction. There is no extra-thoracic extension noted. There is subtle sclerosis of the right anterior aspect of right first rib. The hypermetabolic heterogeneously enhancing soft-tissue lesion

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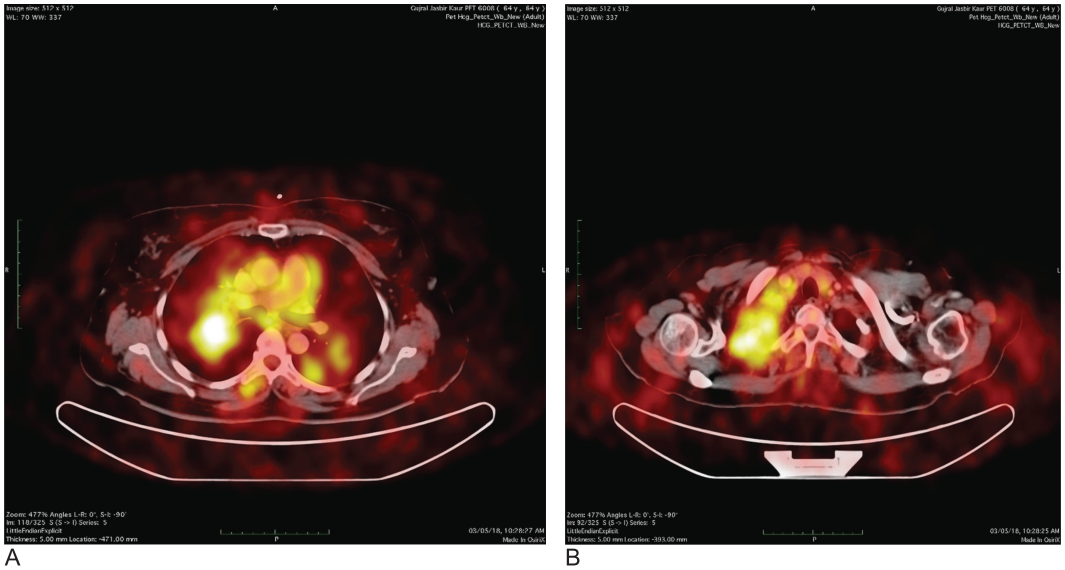


**Figure 1:** The PET-CT scan—a hypermetabolic heterogeneously enhancing soft-tissue mass in the right upper lobe.

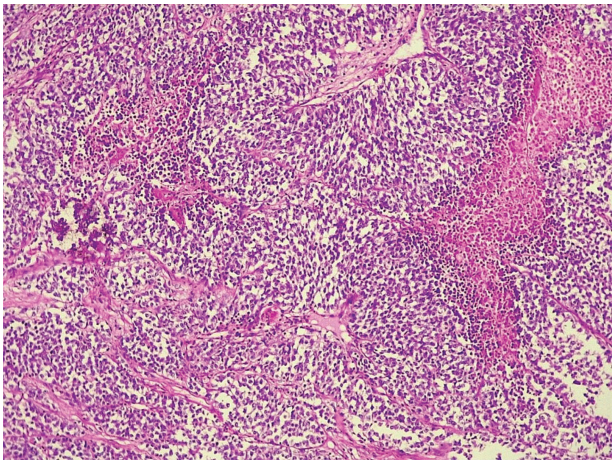

**Figure 2:** H&E 10×: sheets and diffusely spread single lying round to oval undifferentiated cells separated by fibrous septae.

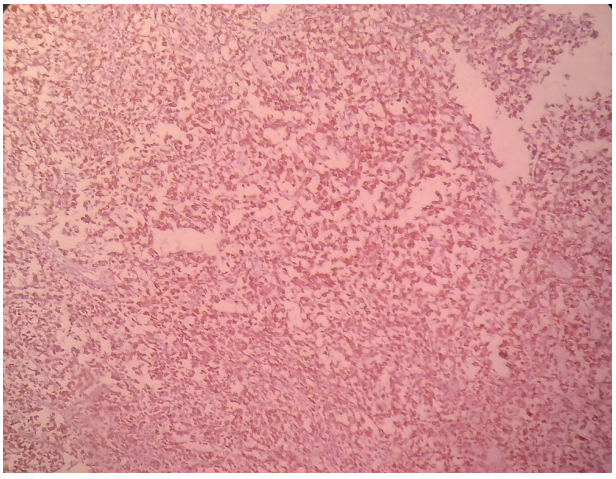

**Figure 3:** Sheets of undifferentiated small blue cells were positive for vimentin.

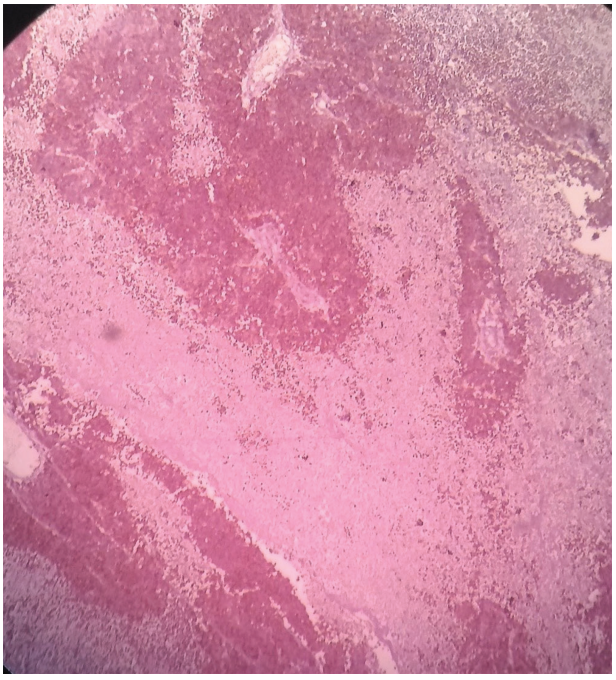

**Figure 4:** Occasional large cell showed cytoplasmic membranous positivity for pan-CK.

in the right upper lobe, its local extent as described above, likely represents second primary neoplastic aetiology rather than metastatic. Mildly hypermetabolic mediastinal nodes are indeterminate. However, the nodal disease cannot be excluded. Hypermetabolic left external iliac and right obturator nodes were suspected for nodal disease. The patient was in good health, and the patient's Eastern Cooperative Oncology Group performance status was good.

A tru-cut image-guided biopsy was performed. The histopathological report was suggestive of metastatic adenocarcinoma. The biopsy of the left external iliac node was performed. Histopath showed reactive lymph node, no malignancy. Right

upper lobectomy was carried out. The post-operative surgical pathology report reiterated the diagnosis of metastatic poorly differentiated adenocarcinoma (Fig. 2). The lesion was hypothesized to be a metastasis of the known primary tumour since the patient was in good general health. The patient had a disease-free survival of more than 3 years. In this case, treatment with a radical intent was warranted. The patient was treated with one cycle of paclitaxel and carboplatin while the paraffin blocks were sent for immunohistochemistry (IHC).

The haematoxylin and eosin (H&E) slides were reviewed before running the IHC markers. The tumour showed two components, one component comprised of small round uniform hyperchromatic blue cells admixed while the other comprised of occasional large cells having central nucleus and moderate cytoplasm. On IHC, the hyperchromatic blue cells were highlighted by vimentin (Fig. 3) and occasional large cells showed immunoreactivity for Pan CK (Fig. 4). Tumour cells showed nuclear beta-catenin positivity. All other markers, CK7, CK20, synaptophysin, CD10, H-caldesmon, EMA, S-100, CD34, SMA, desmin, PLAP, glypican 3, SALL4, CD117 and NUT, were negative in both components. These findings are suggestive of origin from pleuripotent stem cells and ruled out any lineage-specific tumour. Based on the morphology and immunohistochemical findings, a final diagnosis of pulmonary blastoma was made.

The patient was later switched to ifosfamide, carboplatin and etoposide (ICE) chemotherapy protocol. The patient had completed six cycles chemotherapy followed by a PET-CT that showed complete response. The patient was later switched to adjuvant radiation therapy and is on follow up.

## DISCUSSION

Pulmonary blastoma is a rare and aggressive form of lung malignancy. Patients with PPB have a poor prognosis. It is known to grow rapidly and be problematic at the time of diagnosis due to its two-phase localization and construction [2]. PPB is ideally found in children, and its findings in adults are usually rare [2, 3].

In adults, pulmonary blastoma may appear as a large chest mass that may cause cough, pain, hemoptysis and dyspnea. However, in nearly 40% of cases, patients remain asymptomatic. The diagnosis of pulmonary blastoma is often difficult due to its unusual pleomorphic histology. However, it can be suspected when there is cytological evidence of heterogeneity with presence of epithelial and mesenchymal malignant cells [2].

Surgical excision remains the primary choice of treatment for pulmonary blastoma [4, 5]. As per the literature, the prognosis of pulmonary blastoma is poor with a limited survival range of 2–3 years from diagnosis. Differences in survival rate are based on mediastinal node involvement. In two cases, ICE has been used as an effective treatment option for pulmonary blastoma [6, 7]. In a case by Alahwal *et al.* [6], a 3-month complete remission (on follow up) was noted after six cycles of ICE.

The incidence of a second primary synchronous or a metachronous tumour has increased in the past few years by 10%. Based on current evidence, the frequency of the second tumour is estimated to be between 3 and 5%, the third tumour to be 0.5% while the fourth tumour to be 0.3% [8]. Thus, this case is of significant importance for the oncology fraternity as it highlights two aspects, diagnosis of a metachronous tumour and PPB in an adult patient, which is an extremely rare finding. To the best of our knowledge, this is the first case of PPB to be reported in a patient previously diagnosed with endometrial cancer.

## ACKNOWLEDGEMENT

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## CONFLICT OF INTEREST STATEMENT

None declared.

## ETHICAL APPROVAL

No ethical approval was required.

## CONSENT

The patient provided written informed consent for their information and images to be published.

## GUARANTOR

Dr Rajnish Nagarkar.

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