

Classic Biphasic Pulmonary Blastoma in an Adult Patient

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Abstract: *Pulmonary blastoma (PB) is a disease of early childhood. We have reported here a case of classic biphasic pulmonary blastoma (CBPB) in a young adult patient. This is intended at highlighting the fact, that PB occurs in adult individuals, though not a very common occurrence. Thus, high index of suspicion with typically well-defined radiological images may help in clinching diagnosis, though histopathological examination with immunohistochemical evaluation may establish the diagnosis at an early stage, which may be crucial in successful management of such cases.*

Keywords: Pulmonary blastoma, Computed tomography, Immunohistochemistry, Survival, Lung cancer

1. Introduction

Pulmonary blastoma (PB) is a malignant tumor of lung affecting children. PB is a rare entity having an incidence between 0.25-0.5% of all primary lung malignancies. PB occurs exclusively in children younger than 12 years. A 21 year old healthy male, non-smoker was admitted in our hospital with complaint of chest pain, haemoptysis, dyspnea, generalized weakness and loss of weight. On Endobronchial biopsy of left lung mass and immunohistochemistry (IHC) patient was diagnosed with classic biphasic pulmonary blastoma. Patient is not having any significant history of medical illness in the past. Patient is not having family history of similar disease or any other malignancy in 1st and 2nd degree relatives.

2. Future Scope

- PB occurs in adult individuals as well, though not a very common occurrence.
- High index of suspicion with typically well-defined radiological image may help in clinching early diagnosis.
- Histopathological examination with immunohistochemical evaluation may establish the diagnosis at an early stage, which may be crucial in successful management of such cases.

I'm writing this case report, so that my case report can help and guide physicians in faster diagnosis and better management of the patients diagnosed with PB in future.

3. Discussion

Pulmonary blastoma (PB) incidence is 0.25-0.5% of all malignant lung neoplasm and is a rare primary lung malignancy [1]. Due to the histological similarity of the tumor tissue of PB with the fetal lung tissue, Barnard described pulmonary blastoma as "lungembryoma" in 1952,[1]. Till now More than hundred cases of pulmonary blastoma have been reported in literature,[2]. A 21 year old healthy male, non-smoker was admitted in our hospital with complaint of chest pain, haemoptysis, dyspnea, generalized weakness and loss of weight. Patient is not having any significant history of medical illness in the past. At the time of presentation, X-ray of chest Posterior

Anterior view (PA view) was suggestive of well defined round to oval homogeneous radio opacity noted in upper lobe of left lung (fig1). Computed tomography examination of thorax revealed a large well defined rounded in homogenously enhancing soft tissue space occupying lesion in left hilar and suprahilar region with extensions and mass effect (fig 2, 3, 4, 5). Laboratory investigations showed a leucocyte count of 6800/mm³, haemoglobin level of 10.1g/dL, platelet count of 1.5 lac cells/mm³, serum AFP – 2.79ng/ml, serum LDH – 547.3IU/L, serum beta- HCG - < 1.0 IU/ml. Biopsy from left lung mass was taken from endobronchial route, which was histopathologically suggestive of PB (fig 6,7,8). Immunohistochemistry revealed cytokeratin and vimentin positivity (fig 9, 10, 11). Workup for metastasis of the disease was negative. Organ function evaluation like kidney function test (KFT), liver function test (LFT), electrocardiogram (ECG) and echocardiography (2D-ECHO) was done and was found to be within normal limits.

PB is further classified into three different subtypes based on the tissue component:

- A. Monophasic pulmonary blastoma having only epithelial malignant tissue component and it's also known as well differentiated fetal adenocarcinoma (W DFA)
- B. Classic biphasic pulmonary blastoma (CBPB) having both the mesenchymal and epithelial malignant tissue component.
- C. Pleuropulmonaryblastoma (PPB) having only mesenchymal malignant tissue component.

CBPB is most common out of the above mentioned three subtypes,[2]. PPB is consider as an entirely different entity, and is referred to as the pulmonary blastoma of childhood,[3], whereas W DFA and CBPB commonly occurs in an adult individuals,[4]. Since 1999, the World health organization (WHO) classification has separated CBPB from the other two types of PB (W DFA and PPB) where it's now consider as a variant of sarcomatoid carcinomas,[5]. PB is commonly seen in the fourth decade of life, and it has a strong female preponderance,[6]. Our patient, a 21 years old healthy, young male, non smoker

presented with symptoms of chest pain, haemoptysis, dyspnea, and weight loss.

CBPB can occur at any age from birth to seventy years, and out of which 80% of CBPB occurs in an adult individual,[7]. Unlike WDLA and PPB, CBPB is common among the smokers, and it has slight male preponderance,[7]. Symptoms in CBPB are more like an URTI (Upper respiratory tract infection), which require investigations if the symptoms persist, or if it reoccurs. In CBPB, chest X-ray (posterior anterior view) and computed tomography (CT) of thorax is usually helpful and it typically shows a well-defined peripheral or midlung mass,[7]. Immunohistochemistry (IHC) plays a very important role in clinching the diagnosis of CBPB. An endobronchial biopsy from left lung mass was taken in our patient which was histopathologically suggestive of CBPB (fig 6,7,8). IHC revealed vimentin and cytokeratin positivity (fig 9,10,11). Workup for metastasis of the disease was negative. Organ function evaluation like kidney function test (KFT), liver function test (LFT), electrocardiogram (ECG) and echocardiography (2D-ECHO) was done and was found to be within normal limits.

The mainstay of treatment in PB is surgical excision of the tumor with negative margin, though the recurrence rate is quite high even after the resection of the tumor. Adjuvant treatment like post operative chemotherapy and radiotherapy can be quite useful to prolong the patient's survival, but the efficacy of this adjuvant treatment (post operative chemotherapy and radiotherapy) is not yet established and the literature is quite inadequate,[4]. Various chemotherapeutic agents in different combination regimes have been tried as neo-adjuvant and adjuvant treatment,[6, 7].

Due to extensive and inoperable disease, our patient received neo-adjuvant chemotherapy consisting of Ifosphamide and Adriamycin (IE) combination to downstage the tumor. A total of 4 cycles of this combination IE chemotherapy was given to the patient. Toxicities of the above mentioned IE regimen consisted of Grade 2/3 leukopenia during chemotherapy, which were managed successfully. No other hematological, cardiac or urinary toxicity was noted in this patient. Following which, patient was considered for surgery and he underwent left upper lobectomy. Subsequently Patient was given two more cycles of adjuvant chemotherapy comprising of same combination. The factors which point towards the unfavorable prognosis of the patient with PB are the size of tumor more than 5cm, recurrence of the tumor, biphasic type on IHC, metastatic disease at the time of presentation, and the frequent lymph node involvement,[1, 2]. In our patient, on the basis of the presence of two important prognostic factors the biphasic pattern of the disease on IHC and the large size of the tumor on X-ray chest (PA view) and computed tomography of thorax, patient was planned for adjuvant chemotherapy following resection of the tumor. Although the long term survival of the patient with PB have been reported in several individual case reports, but the overall prognosis of the patient with PB remain to be poor,[8]. In

one survey of 83 patients with PB, approximately mean survival of 33 months was reported in those patients where negative margin was achieved during surgical excision of the tumor, [8].

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Figure Captions

Figure 1: Well defined round to oval homogeneous radioopacity is noted in upper lobe of left lung

Figure 2 Fig 3 Fig 4 Fig 5 : Well defined round to oval heterogenous soft tissue attenuating mass is noted in apicoposterior segment of left upper lobe with extension into apical segment of left lower lobe

Figure 6: Tumor bits lined by cubo-columnar epithelial cells with focal Pseudostratification

Figure 7: The mesenchymal cells show round to spindle shaped hyperchromatic nuclei, inconspicuous nucleoli and indistinct cytoplasm.

Figure 8: Interspersed areas of necrosis.

Figure 9: The lining epithelial cells are positive for cytokeratin while the Mesenchymal cells are negative (X200 cytokeratin).

Figure 10: The lining epithelial cells are positive for cytokeratin while the Mesenchymal cells are negative (X400 cytokeratin).

Figure 11: The mesenchymal cells show immunopositivity for vimentin

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Figure 1

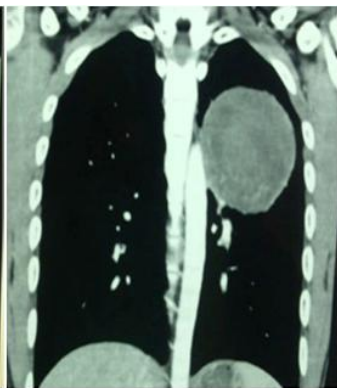


Figure 2



Figure 3



Figure 4

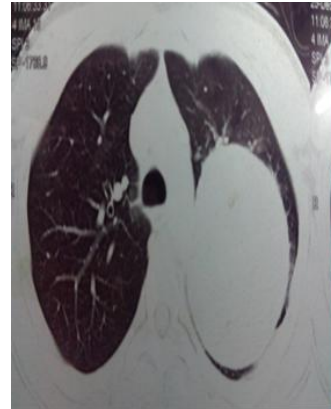


Figure 5

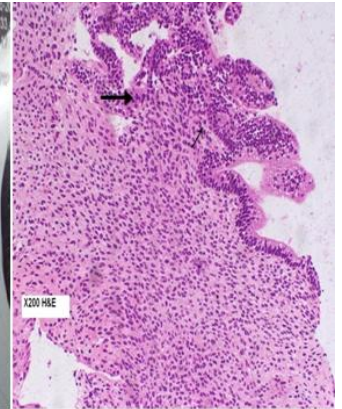


Figure 6

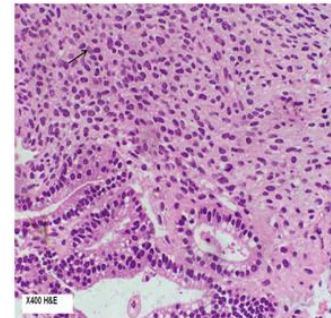


Figure 7

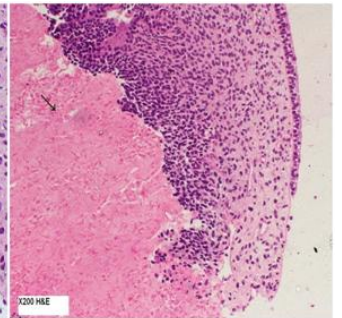


Figure 8

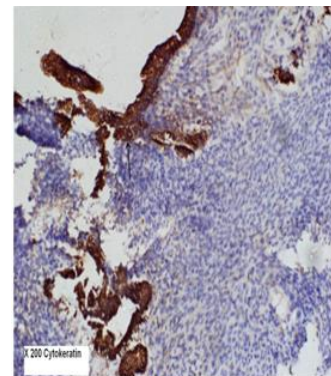


Figure 9

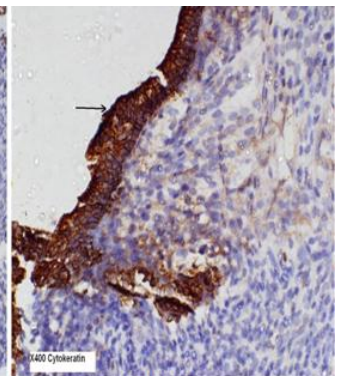


Figure 10

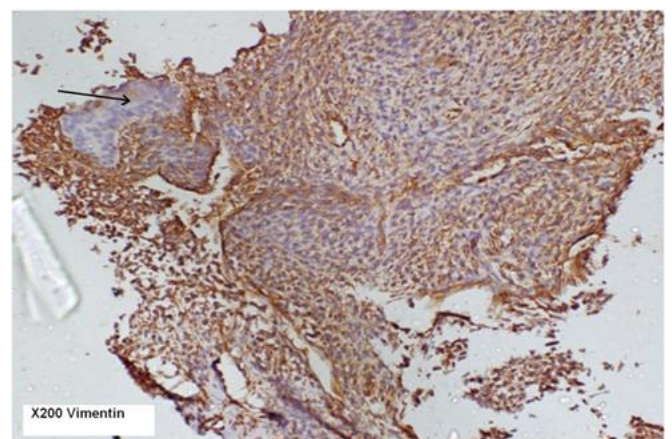


Figure 11