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Pulmonary Blastoma with Focal Yolk Sac Differentiation- A Diagnostic Challenge

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Abstract

Background: *Pulmonary blastoma is an unusual primary lung malignancy, constituting about 0.25%- 0.5 % of all primary lung tumors. It is asymptomatic in up to 40% of patients. Cough, chest pain, hemoptysis, fever, and dyspnoea were common symptoms. Clinically diagnosis may be difficult so, histopathology is necessary to confirm the diagnosis.* ***Case presentation:*** *We present a case of 14 years male child presenting with fever, cough, chest pain, breathlessness and occasional blood in the cough. Clinically and radiologically the lesion was diagnosed as pneumonia. The patient died despite treatment. An autopsy was performed. The diagnosis was done on histopathology of lung tumor as pulmonary blastoma immunohistochemistry showed focal immunoreactive result with Glypican-3 and alfa Fetoprotein.* ***Conclusion:*** *because of the rarity of pulmonary blastoma and to aware recent WHO reclassification of the lung tumor, we reported this case. Now pulmonary blastoma is classified as a separate entity and subtype of sarcomatoid carcinoma which is non-small cell carcinoma. It should be differentiated from fetal adenocarcinoma, pleuro-pulmonary blastoma.*

Keywords*: Pulmonary blastoma, Yolk sac tumor, pleuro-pulmonary blastoma, autopsy*

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Introduction

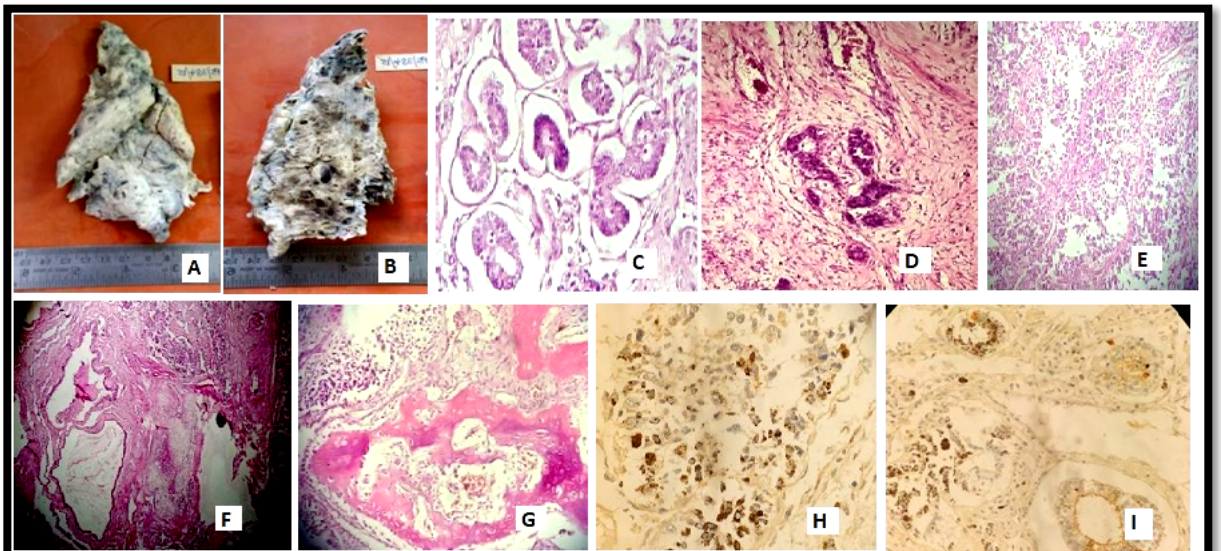
Pulmonary blastoma is a rare primary aggressive pulmonary tumor accounting for less than 1% of all primary lung tumors. [1-6] According to WHO classification [2015] pulmonary blastoma is considered as one of the subtypes of sarcomatoid carcinoma which is a non-small cell carcinoma of the lung. Morphologically pulmonary blastoma is a classic biphasic tumor that consists of well-differentiated fetal adenocarcinoma and primitive mesenchymal stroma including undifferentiated blastemal elements [6]. This tumor commonly occurs in the pediatric age group and also reported in adults [2].

Case History

A 14 years male child admitted to the hospital for complaints of fever, cough, chest pain, breathlessness for 1 month. Breathlessness was insidious in onset, gradually progressive and more on exertion. He also complained of occasional blood in cough, reduced appetite, decreased weight and fatigue. USG thorax revealed a large mass with multiple cystic areas involving almost whole left lung and minimally increased pleural fluid noted on the left side. He was clinically and radiologically diagnosed as pneumonia and was treated symptomatically. On Clinical & Radiological examination, a diagnosis of lobar pneumonia of left Lung with pleural effusion was made. The patient was treated with antibiotics and symptomatic drugs.

Hematological investigations revealed hemoglobin was 8 gm%, WBC count was neutrophilic leucocytosis, with normal platelet count. Biochemical investigations were unremarkable. The general condition of the patient deteriorated and he died on the 8th day of admission. A clinical postmortem was performed. On gross examination, the upper lobe of the left lung measured 15 x 7 x 5cm. External & cut surfaces showed a solid, grey-white diffuse firm mass with multiple small cysts involving almost the whole upper lobe of the left lung, with few hemorrhagic areas. Pleura was thickened and opaque. The right lung was normal. Grossly, the brain and all abdominal organs were unremarkable.

Histopathologically, multiple sections from the left lung mass revealed a tumor consisting of a classic biphasic pattern showing epithelial and mesenchymal malignant components. The epithelial component comprised of well-differentiated adenocarcinoma of fetal type with focal yolk sac differentiation. The mesenchymal element comprised of small round undifferentiated blastematous cells with vesicular nuclei and scanty cytoplasm arranged in sheets, nests or clusters. There were foci of mesenchymal differentiation into osteogenic, chondrogenic elements without malignant change. Increased mitotic activity and areas of hemorrhages and necrosis were noted. The tumor was seen infiltrating into the pleura. This tumor consisted of a classic biphasic pattern of well-differentiated adenocarcinoma and primitive undifferentiated blastemal cells which is a malignant mesenchymal component. So it was diagnosed as pulmonary blastoma with yolk sac differentiation. The right lung showed congestion with no evidence of tumor. Other organs including the brain, liver, spleen, kidneys, adrenals GIT were unremarkable.



A & B: External and cut surfaces of left lung tumor. **C**: Epithelial element showing well differentiated adenocarcinoma. **D**: Mesenchyme. **E**: Primitive mesenchymal component showing undifferentiated blastemal cells[H& E STAIN]. **F & G**: Cystic changes and osseous metaplasia. **H & I**: Glypican-3 Positive and alpha-fetoprotein focally positive.

Discussion

Pulmonary blastomas are a rare aggressive neoplasm comprising 0.25-0.5% of all primary lung tumors [7]. Morphologically they mimic fetal lung tissue before 4 months gestation [8]. First described by Barnard in 1952 [9], they have since been divided into three subgroups: classic biphasic pulmonary blastoma, well-differentiated fetal adenocarcinoma also called monophasic pulmonary blastoma- and pleuropulmonary blastoma of childhood. While well-differentiated fetal adenocarcinoma contains malignant glands and benign-appearing mesenchymal tissue and pleuropulmonary blastoma contains malignant glands of embryonal appearance and benign-appearing epithelium, classic biphasic pulmonary blastoma

contains glands and mesenchymal tissue that are both embryonal and malignant. In the 1999 and 2004 WHO classifications [10, 11], well-differentiated fetal adenocarcinomas and pleuropulmonary blastomas were separated from the biphasic tumors. Classic biphasic pulmonary blastoma is now considered as part of the spectrum of sarcomatoid carcinomas. It typically presents with cough, hemoptysis, dyspnea or chest pain due to tumor impinging on the bronchi or pleura. Forty percent of cases may be asymptomatic [12]. The average age at diagnosis is 40 years with an increased frequency in males [2:1] [13]. The clinical examination may reveal a localized reduction in breath sounds or sequelae of cigarette smoking with over 80% of cases associated with a smoking history. Abnormalities in laboratory tests are infrequent and non-specific. Pulmonary blastoma almost always presents as a unilateral, large, well-circumscribed, solitary mass on chest radiograph. Given the often, peripheral nature of these tumors, tissue diagnosis by bronchoscopy only occurs in 25% of cases [12] but they can often be visualized on thoracic ultrasound with findings correlating well with those seen on CT [14]. The role of CT in the radiological diagnosis of pulmonary blastoma is difficult. With our case demonstrating final pathological is inconsistent with clinical and radiological diagnosis. Due to the challenging nature of the histology, a preoperative diagnosis is only obtained in one-third of cases [7] Differential diagnoses must include benign conditions such as hamartoma and pleural fibroma as well as malignant conditions such as other primary or metastatic lung cancers. Pulmonary blastomas are biphasic tumors that are part of the sarcomatoid carcinoma subgroup [10], which also include carcinosarcomas [defined as a malignant tumor having a mixture of carcinoma- and sarcoma- containing heterologous elements such as malignant cartilage, bone, or skeletal muscle] and pleomorphic carcinomas [similar tumor without heterologous elements], both histologically resembling adult-type carcinomas and sarcomas. The epithelial component of classic biphasic blastoma is composed of tubules of glycogen-rich, non-ciliated cells that resemble fetal lung of the pseudo-glandular stage of lung development with sub-nuclear and supra-nuclear glycogen vacuoles. The embryonic appearance of the stroma is due to the small size, oval and spindle shape of the cells, and myxoid matrix. Classically this blastematous stroma does not express cytokeratins or pulmonary markers. Beta-catenin may play a role in tumorigenesis of classic pulmonary blastoma. Its aberrant nuclear/cytoplasmic localization by immunostaining has been reported to be useful in distinguishing classic pulmonary blastoma from a blastomatoid variant of carcinosarcoma and high-grade fetal type adenocarcinomas [15]. On IHC study as Glypican-3 and Alpha-fetoprotein were focally positive in this tumor so this confirms the final diagnosis of this lung tumor as pulmonary blastoma with focal yolk sac differentiation.

Surgery is the optimal treatment for localized disease. Mean survival of 33 months was reported in resected cases compared to 2 months in those with the un-resected disease. Limited resections do better than pneumonectomies [13], presumably due to less extensive tumor burden. Larsen [13] reported a 16% response rate into chemotherapy in 43 cases of classic biphasic pulmonary blastoma. No agent is known to be more effective than another, but cisplatin is often used given its efficacy with primitive tumors. Most cases have not shown a response to radiotherapy. Prognosis is poor with two-thirds of patients dying within 2 years and only a 16% 5-year survival. Prognosis is determined by the size of the tumor at the time of diagnosis, with tumors <5 cm doing better. Tumor metastasis and tumor recurrence despite resection both predict a poor prognosis. Unfortunately, 43% of tumors recur within 1 year with a propensity for sites such as brain and mediastinum [12]. Recurrence tends to occur within 1 year after diagnosis or not at all [13].

Conclusion

As pulmonary blastoma is a very rare entity, confirmatory diagnosis is not possible without histopathological examination. We present a case of pulmonary blastoma in a14 years male child which was not diagnosed. It is necessary to create awareness of newer WHO reclassification of the pulmonary tumors which has mentioned pulmonary blastoma is as a separate lung tumor. surgical intervention is the choice of treatment with adjuvant radiotherapy and chemotherapy as

per requirement. A central registry of individual experience may be beneficial to improve our knowledge of uncommon lung tumors.

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