



Biphasic Pulmonary Blastoma: a Case Report and Literature Review

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Authors' contributions

This work was carried out in collaboration between all authors. Author TS was involved in the analysis of the data and the literature search and wrote the manuscript. Author IO helped with the patient management and revision of the manuscript. Authors AM and SB helped with the literature research. Authors YB and HM helped with modifications and revision of the manuscript. Author HE approved the treatment and analyzed the literature data. All authors read and approved the final manuscript.

Case Study

Received 26th March 2013
Accepted 4th May 2013
Published 24th May 2013

ABSTRACT

Introduction: Biphasic Pulmonary blastoma (BPB) is classified as one of the rare primary lung malignancies. It is composed of a mixture of epithelial and mesenchymal tissues resembling embryonic lung tissue. BPB is considered to be distinct from other lung tumors based on pathological features, clinical course and prognosis.

Presentation of Case: The authors report an atypical case of BPB in a 27 -year-old man presented with complaints of dyspnea and left-sided chest pain for the previous four months. A chest radiograph showed the presence of an opaque left hemithorax, and the mediastinum was pushed toward the left. Computed tomography (CT) of the chest revealed a mixed solid and cystic process with variable contrast enhancement measuring 15,4 x 13,7 cm occupying the totality of the left hemithorax, pleural effusion, and a collapsed left lung, with contralateral mediastinal shift. A transthoracic needle pleural biopsy yielded a diagnosis of BPB. A general examination shows a peritoneal effusion. One month after diagnosis, the tumor grew rapidly, and therefore he was treated only by palliative care. He died from respiratory failure one month later. Although BPB is rare, this entity is increasingly described.

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Conclusion: The purpose of presenting this case report is to raise awareness among clinicians to consider this clinical entity as a differential diagnosis when a pleural mass is identified. Histological examination is the most reliable and conclusive method of diagnosing BPB and differentiating it from other primary or metastatic lung malignancies.

Keywords: Biphasic; pulmonary; blastoma.

ABBREVIATIONS

BPB: Biphasic Pulmonary blastoma; CT: Computed tomography; IHC: immunohistochemistry.

1. INTRODUCTION

Biphasic pulmonary blastoma (BPB) is a rare primary lung malignancy [1]. It is characterized by primitive blastoma and malign mesenchymal stroma which presents multiple differentiation in terms of histology. It is considered to be distinct from other lung tumors based on pathological features, clinical course, and prognosis. Due to its rarity, there is currently no treatment guideline, especially concerning the roles of chemotherapy and radiotherapy. In this report, a rare BPB exhibiting rapid growth and fatal outcome is described. The diagnostic approach to our patient is presented and the pertinent literature is reviewed.

2. CASE PRESENTATION

A 27 -year-old man presented with complaints of dyspnea and left-sided chest pain for the previous four months. He had an 8-year history of tobacco use, smoking one half pack of cigarettes per day for 8 years, but had no additional risk factors. During a clinical examination, the breath sounds were decreased on the left side. A chest radiograph showed the presence of an opaque left hemithorax, and the mediastinum was pushed toward the left (Fig. 1). Computed tomography (CT) of the chest revealed a mixed solid and cystic process with variable contrast enhancement measuring 15,4 x 13,7 cm occupying the totality of the left hemithorax, pleural effusion, and a collapsed left lung, with contralateral mediastinal shift (Fig. 2). Bronchoscopic examination was evaluated to be nondiagnostic. An interventional radiologist performed a transthoracic needle pleural biopsy. Histologically, the tumor was composed of biphasic pattern comprising ductal structures and stromal components comprising spindle cells. The interstitium was composed of pleomorphic and spindle cells; mesenchymal component suggestive of rhabdomyosarcoma was identified (Fig. 3). After the pathological diagnosis, a CT scan of the abdomen was performed, which revealed a peritoneal effusion. However, the tumor grew rapidly, with pleural effusion. Our patient was treated only by palliative care. He died one month later due to acute respiratory failure.



Fig. 1. chest radiograph showed an opaque left hemithorax, and the mediastinum was pushed to the left



Fig. 2. CT of the chest revealed a mixed process occupying the totality of the left hemithorax, and a collapsed left lung, with contralateral mediastinal shift

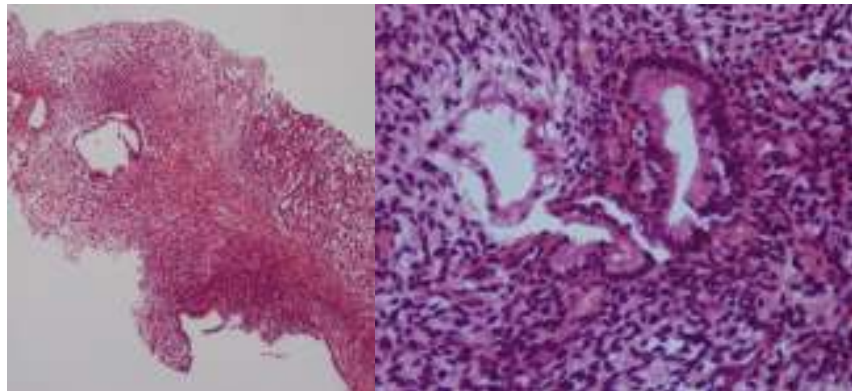


Fig. 3. Histological features of the lesion containing epithelial and mesenchymal component consistent with pulmonary blastoma : 20X(right)40X(left).

3. DISCUSSION

BPB is classified as one of the rare primary lung malignancy, comprising only 0.25–0.5% of all malignant lung neoplasm [1]. Initially, Barnard described it as “lung embryoma” in 1952; the term blastoma was introduced later in 1961 by Spencer to show that these tumors arising from the pulmonary blastema and other tumours developing from fetal tissue appears to share the same histological features [2,3]. More than one hundred cases have been reported in literature. To the best of our knowledge, no large series has documented the exact number of BPB cases worldwide. Data are available in isolated case reports and small case series.

BPB have been subdivided by Koss et al. [4] into three subgroups: well-differentiated fetal adenocarcinoma comprising epithelial malignant component only, pleuropulmonary blastoma of childhood showing features of mesenchymal malignant components only and classic BPB characterized by both epithelial and mesenchymal malignant components, which is the most common of these three subgroups.

Approximately 80% of BPB occurs in adults, one peak in age is described in the fourth decade [4]. There is a slight male preponderance and is common among smokers [4].

Clinically, BPB presents with non-specific respiratory symptoms that may mimic an upper respiratory infection (cough, haemoptysis, dyspnoea and chest pain). However, 40% of patients are asymptomatic and found coincidentally on chest radiography, which shows usually a well-circumscribed peripheral or central mass with no definite lobar predominance [5], in our case, chest radiograph showed the presence of an opaque left hemithorax, and the mediastinum was pushed toward the left.

On CT, BPB presents as a heterogeneous mixed (solid and cystic) lesion with variable contrast enhancement with a necrotic centre [6]. The presence of pleural or mediastinal involvement is an indication of metastatic disease.

Preoperative diagnosis of pulmonary blastoma is difficult because the expansive tumor growth in the bronchial tube interferes with transbronchial diagnosis. On reviewing the literature; we found that most of the cases were diagnosed by surgical biopsy; in the third of cases the diagnosis has been made by bronchoscopy or CT-guided transthoracic biopsy.

Histological examination supplemented with IHC staining studies is the most reliable and conclusive method of diagnosing BPB and differentiating it from other more frequent primary lung malignancies. Microscopically, BPB is characterized by the presence of two or biphasic malignant and immature cellular components: epithelial and mesenchymal.

The mesenchymal cellular component is represented by round or oval, rarely fusiform, shaped cells, with a high mitotic index and scattered in a myxoid stroma. It can demonstrate rhabdomyoblastic (with desmin and myogenin expression in immunohistochemistry), chondroid or osteoid differentiation [7]. The epithelial component consists of glandular structures with columnar cells. The p53 protein is generally overexpressed [8]. In our case, microscopic examination confirmed the biphasic pattern comprising epithelial and malignant mesenchymal components consistent with BPB, the immunohistochemistry was not necessary.

BPB is an aggressive tumour associated with a poor prognosis. As a consequence of the rarity of this disease, there is no recommendation for the treatment of such malignancy. In

limited disease, surgery is the cornerstone of curative intent treatment. The efficacy of adjuvant chemotherapy and radiotherapy is still debated.

In advanced disease, there is no standard chemotherapy regimen. Doxorubicin plus ifosfamide, cyclophosphamide, vincristine, VP-16 and cisplatin plus etoposide provided objective response [9,10]. Although, several individual case reports of long-term survival have been documented, the overall prognosis of this rare malignancy is poor, and two-thirds of patients die within 2 years of diagnosis [11]. The factors contributing to the unfavorable prognosis are the biphasic type, tumor recurrence, metastatic disease on presentation, tumor size over 5 cm, and frequent lymph node involvement [1,12].

In our case factors of unfavorable prognosis were the biphasic type and tumor size over 5 cm, the diagnosis of BPB was made later, after deterioration of the general condition of our patient and aggressive progression of the tumor, which was unresectable.

4. CONCLUSION

BPB is a rare tumor, and many of its features remain unsolved [13]. Thus, clinical data of BPB patients need to be accumulated. Since no standard treatment is currently available, treatment should be carefully assessed on a case-by-case basis.

CONSENT

Not applicable.

ETHICAL APPROVAL

Not applicable.

COMPETING INTERESTS

The authors declare that they have no competing interests.

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Peer-review history:
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