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Case Report
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Biphasic Pulmonary Blastoma - A Rare Lung Tumor: Case Report

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Abstract

Pulmonary blastoma is a very rare neoplasm, similar to fetal lung tissues, comprising less than 0.5% of all primary lung tumors. The mainstay of treatment is surgical resection but, due the rarity of this tumor, remains unclear the role of adjuvant therapy (radiation or chemotherapy). The prognosis of this malignancy is poor and the overall 5-year survival is around 15%. We report a case of a 51 year-old woman who presented with cough, haemoptysis and a tumor shadow in the right middle field of a chest radiography. The pathological diagnosis of biphasic pulmonary blastoma was made after surgical resection of the tumor, and the patient received an adjuvant platinum based chemotherapy. The patient is in complete remission and currently on a two year ongoing follow-up.

Introduction

Pulmonary blastoma is a very rare neoplasm, similar to fetal lung tissues, comprising 0.25% to 0.5% of all primary lung tumors [1]. In 1945, Barret and Barnard reported this disease for the first time and in 1952, Barnard called it "embryoma of the lung" [2,3]. The name "pulmonary blastoma" was suggested by Spencer in 1961 [4]. Initially pulmonary blastoma was classified into three subtypes based on tissue component: 1) the classical biphasic Pulmonary Blastoma (PB), characterized by both epithelial and mesenchymal malignant components, 2) Well-Differentiated Fetal Adenocarcinoma (WDFA) contains epithelial malignant component only, and 3) Pleuropulmonary Blastoma (PPB), a pediatric tumor comprising the mesenchymal tissues [1,5,6].

However, since 1999, that the World Health Organization (WHO) classification of lung tumors classifies the PB as carcinoma with pleomorphic sarcomatoid or sarcomatous element, the WDFA is included as an histological variant of adenocarcinoma and the PPB is considered a pediatric soft tissue tumor, occuring in children younger than 6 years old [7].

Histogenesis of the PB is yet unknown and the risk factors are not clearly defined, but a strong association with smoking has been suggested [6,8]. Usually without sex predominance, most recent data suggest a slightly male predominance and a peak of incidence in the 4th and 7th decade of life [6].

Patients with PB can present with an incidental finding of a lung mass on chest radiography (40%) or with non-specific respiratory symptoms, like cough, dyspnea, hemoptysis and chest pain [5,9]. Metastatic cancer is frequent and is typically found in the brain, mediastinum, pleura, diaphragm and liver [1,6].

Surgery is the mainstay of treatment and the prognosis is poor with a 5-year survival rate of 16% to 25% and a 10-year of 8% [6,8].

Case Presentation

We present a case of a 51 year old female, melanodermic, natural from Cape Verde, smoker (25 pack-year) with no other relevant past medical history or usual medication. Evacuated to Portugal due to persistent cough and haemoptysis, with no constitutional symptoms or other complaints. Blood tests showed negative serologies and bacilloscopy, and chest X-ray showed a nodular lesion in the third middle of the right lung. The chest Computed Tomography (CT) showed an hypodense well-demarcated mass of the Upper Right Lung (URL) lobe with approximately 4 cm, without contrast capture and without evidence of significant lymphadenopathy or pleural effusion (Figure 1). A flexible bronchoscopy revealed a subtotal occlusion of the most posterior subsegmentar bronchus of B3 by pedunculated and hemorrhagic lesion. Bronchoalveolar lavage and lung biopsies were negative for neoplastic cells. The investigation was completed by a

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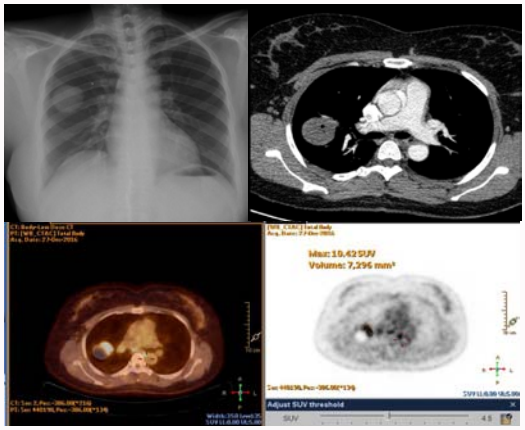

Figure 1: (A) Chest X-ray with a right well-rounded pulmonary opacity. **(B)** Computed tomography scan showing an hypodense well-demarcated 4 cm mass of the right upper lobe, without contrast capture. **(C and D)** Computed tomography/positron emission tomography (CT-PET) revealing an abnormal 18F-FDG uptake around the periphery of the lesion (SUV 10,4) and in the ipsilateral hilar lymph node (SUV 5).

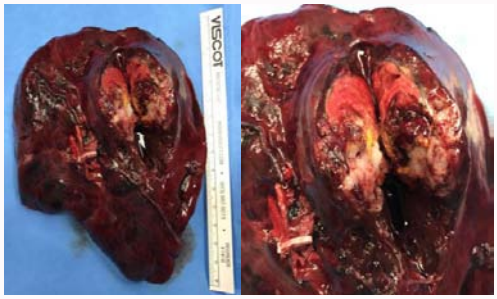

Figure 2: A right posterolateral thoracotomy with a upper lobectomy. **(A)** Macroscopic vision of the tumor infiltrating the RUL. **(B)** The cutting surface of the heterogeneous tumor.

Computed Tomography/Positron Emission Tomography (CT/PET) showing a hypercaptation at the periphery of the known URL lesion, with SUV_{max} of 10.4, a significant part of the lesion contents seems to be liquid-necrosis, cystic lesion? right hilar adenopathy with SUV of 5 and hypermetabolic right paravertebral adenopathy (Figure 1). Pre-operative work-up showed no contraindications and the patient underwent surgery. The patient underwent an upper right lobectomy and mediastinal node dissection by posterior thoracotomy (Figure 2). The postoperative period was uneventful, the drains were removed on 4th day after surgery and the patient was discharged one day later. Anatomopathological examination revealed a biphasic pulmonary blastoma staged as pT2aN0. After multidisciplinary discussion, it was decided to proceed with adjuvant platinum based chemotherapy. The patient received four cycles of Cisplatin and Etoposide with tolerance and significant clinical improvement. The patient is in complete remission and currently on a 2 year ongoing follow-up.

Discussion

Pulmonary blastoma is a rare, highly aggressive and poorly differentiated neoplasm. Since the first publication of this malignancy, in 1945, only a few hundred cases have been described in literature. Despite the fact that no risk factor has been determined, smoking is reported in almost 80% of the cases [6].

There are no specific symptoms of PB and 40% of the cases are asymptomatic, which causes the delay in the diagnosis. The preoperative pathological diagnosis by bronchoscopy or CT guided biopsy may be difficult to obtain because of the unusual pleomorphic histology of this tumor [1]. As happened with this patient, in majority of the cases the surgery plays both, a diagnostic and curative role.

Although, the literature shows a several individual case reports of long-term survival, the overall prognosis of biphasic pulmonary blastoma is really poor and 60% of the patients die within 2 years [6]. Contrary to our clinical case, the presence of metastasis at the time of diagnosis is common (~43% of the cases) and usually affects the brain, mediastinum, pleura, diaphragm and liver [10]. Local recurrence is common and tends to occur within the first 12 months after surgical resection. Tumor recurrence, presence of metastasis, lymph node involvement and tumor size over 5 cm were considered to be un favourable prognosis factors [6,11].

Complete surgical excision is the treatment of choice for PB. Radiotherapy and chemotherapy have been described as an alternative or adjuvant treatment but, due the rarity of this tumor, its efficacy and prognostic impact is not clear. Nevertheless, several case reports described the use of platinum-based adjuvant chemotherapy [12].

Finally, we conclude that pulmonary blastoma is a rare primary lung malignancy with poor prognosis. Symptoms are nonspecific and most are incidentally discovered on routine radiography. Preoperative diagnosis may be difficult to obtain because of the unusual pleomorphic histology. There are no clearly defined treatment regimens, although complete surgical resection appears to confer the best prognosis.

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