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Case Report Open Access

Pulmonary Blastoma in an adult male

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Introduction

Pulmonary blastoma is one of the rare lung tumors and is considered to be distinct from other lung tumors by its pathological features, clinical course and prognosis [1]. Classic pulmonary blastoma is composed of both malignant mesenchymal stroma and epithelial components resembling embryonic lung tissue. Surgery is the standard treatment and the efficacy of adjuvant chemotherapy and radiotherapy has not yet been established [2].

Case Report

Our patient was admitted and managed at Abassia Pulmonary Hospital, Ministry of Health, Cairo Governorate, Egypt. A 37-year-old Egyptian male presented to our hospital after repeated attacks of blood tinged sputum. The first attack dated about 6 months before admission. The patient had progressive shortness of breath over a period of two years. The patient was a smoker of one pack of cigarettes per day for more than 15 years. There was no significant past history nor family history. After admission to the Chest Medicine Department at Abbasia Pulmonary Hospital, routine basic investigations were done including complete blood picture with differential count, fasting blood sugar and 2 hours after meal, liver function tests, renal function tests, and coagulation profile. All investigations were within normal. Sputum examinations for AFB for 3 successive days were done and were all negative. Sputum cytology did not show malignant cells. The P-A view of his chest X-ray showed a well-demarcated right upper and middle lung zones mass. The mass extends to the right lung apex

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abutting the inner chest wall and medially extended to the mediastinum and the right hilum. The mass has a heterogeneous texture (Figure 1). Chest CT scan (Figure 2) showed a large well defined, heterogeneous right upper lobe mass. The CT showed no significant meditational lymph nodes enlargement. Preoperative fiber optic bronchoscoy showed no endobroncheal pathology. Also, the broncho-alveolar lavage was negative for both AFB and malignant cells. Preoperative CT needle guided biopsy revealed a biphasic pulmonary blastoma with both epithelial and mesenchymal components. Preoperative CT abdomen and bone scintigraphy revealed no evidence of distant metastasis. The patient was referred to the Thoracic Surgery Department at our hospital for resection of the pulmonary blastoma affecting the right upper lung lobe. General anesthesia was conducted via double lumen endotracheal tube for one lung anesthesia. The patient was positioned in the lateral decubitus position with his right side up. Standard right postero-lateral thoracotomy in the fifth space was done. The tumor mass was seen and felt involving the entire right upper lung lobe and sparing both the middle and lower lung lobes. The mass was well circumscribed, lobulated with firm to hard texture. Classic right upper lobectomy was done. After closure of the bronchial stump with simple interrupted Vicryl 2^0 sutures, the stump was covered using a pedicled intercostal muscle flap as our standard technique in cases of malignancy and TB to guard against postoperative bronch-pleural fistula.

Postoperative pathology report confirmed the diagnosis of high grade biphasic pulmonary blastoma with free bronchial margin, free pleural tissue and negative mediastinal lymph nodes for metastasis. The postoperative course of the patient was uneventful. After removal of the stitches; the patient was referred to the Cancer Institute of Cairo University for evaluation and the possibility of post lobectomy chemotherapy and / or radiotherapy.

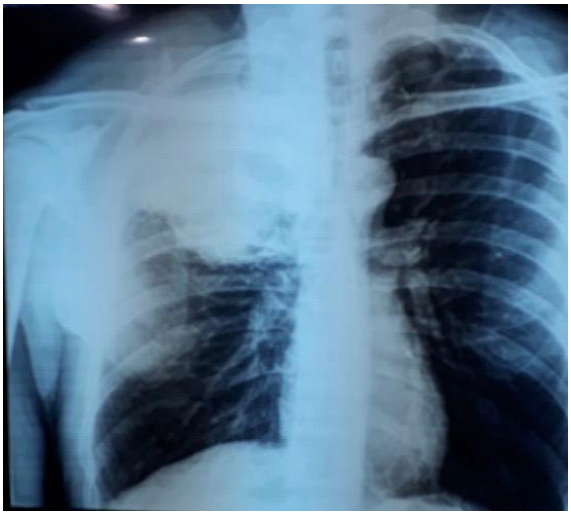

Figure 1: Chest X-ray P-A view showing.

Right upper and mid zonal well-defined tumor mass.

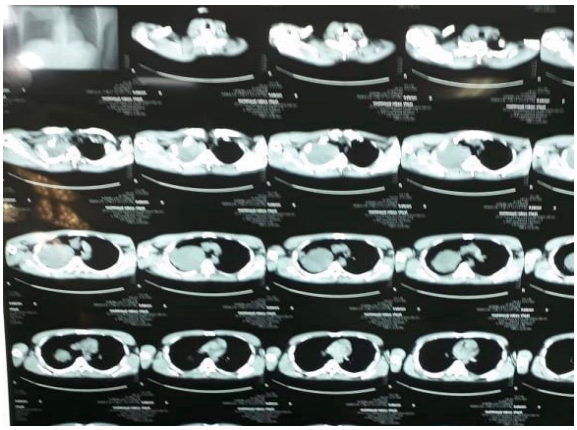

Figure 2: Mediastinal window of CT.

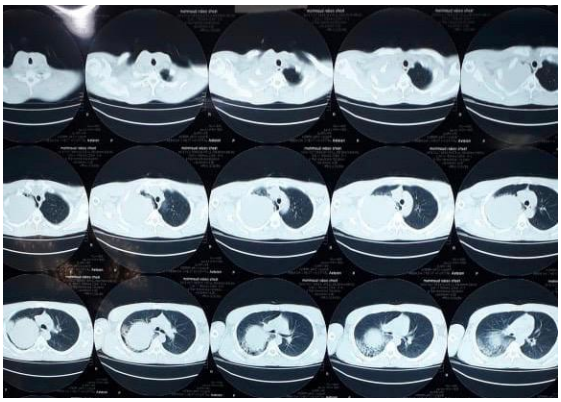

Figure 3: Pulmonary window of CT chest chest showing right upper lobe mass. showing the right upper lobe tumor mass.

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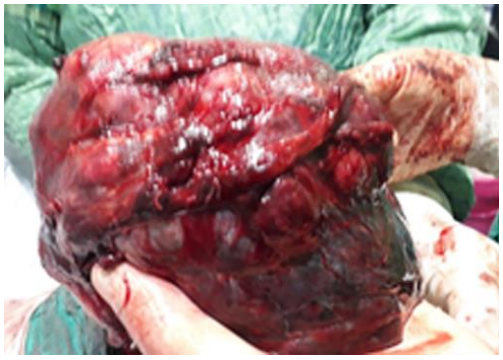

Figure 4: The right upper lobe.

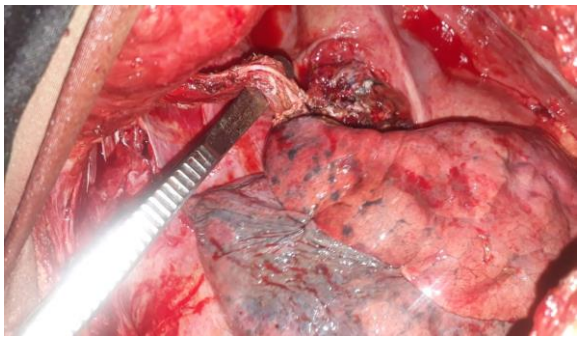

Figure 5: Intercostal muscle flap covering lobulated tumor mass. the bronchial stump.

Discussion

Pulmonary blastoma is a rare primary lung tumor occurring mainly in adults. The incidence is 0.25 to 0.5 % of all primary lung tumors [3]. The tumor was first described by Barrett and Barnard in 1945, and it was termed by Barnard as "embryoma" [4]. The term "blastoma" was suggested by Spencer in 1961 because the pathogenesis of that tumor was believed to be similar to Wilms' tumor or nephroblastoma [5]. The World Health Organization (WHO) had classified pulmonary blastomas into three categories according to the histopathology, the first type is pulmonary blastoma which is biphasic, the second type is pleuropulmonary blastoma which is monophasic and occurs commonly in childhood and the third type is the well differentiated fetal adenocarcinoma (WDFA) [6].

The etiology and predisposing factors are not still fully understood. But, in the literature, there is strong evidence showing the correlation between cigarette smoking and pulmonary blastoma [7]. Our patient was a smoker of one pack per day for more than 15 years. The peak incidence of biphasic pulmonary blastoma is in the fourth decade of life. About 30% of cases occurring in children and the tumor is known as pleuropulmonary blastoma [8]. The age of our patient at the time of presentation was 37 years old.

Some groups reported a male predominance while others did not [9]. Approximately 60% of patients are symptomatic [10]. Usually biphasic pulmonary blastoma are located in the periphery of the lung, while 25% present with end bronchial tumor [11]. Our patient was a male who complained of blood tinged sputum and the tumor was proved by pre and postoperative histopathology to be a biphasic peripheral pulmonary blastoma. The radiological appearance of pulmonary blastoma is usually a solitary pulmonary nodule or a mass with a smooth margin due to the desmoplastic changes surrounding the tumor. Rarely the tumor cavitates, calcified or be multiple [12].

Preoperative tissue diagnostic procedures e.g. bronchoscopy, mediastinoscopy, CT needle guided biopsy usually cannot diagnose pulmonary blastome due to lack of cellular material and extensive necrosis of the tumor [13]. In our patient CT guided biopsy could diagnose the tumor to be biphasic blastoma and this histopathology was confirmed by the postoperative histopatopathological examination. The standard treatment of pulmonary blastoma consists of surgical resection [14]. Our patient had a resectable tumor and a right upper lobectomy was performed. Due to the lack of data, adjunctive therapy with chemotherapy and / or radiotherapy is still controversial [9]. Our patient was referred to Cancer Institute of Cairo university for further evaluation of his need for adjunctive therapy or not. The prognosis of pulmonary blastoma is poor regardless of the histology. The overall 5 years survival was reported to be 16 % [15]. Another study the researchers had reported that biphasic pulmonary blastomas have a poorer prognosis

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than the monophasic well differentiated fetal adenocarcinoma which have a better prognosis and a five years survival around 50% [16]. The factors contributing to the unfavorable prognosis are the biphasic type, tumor recurrence, metastatic disease on presentation, tumor size more than 5 cm, and frequent lymph node involvement [17].

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