Case Report

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Pulmonary blastoma: a case report of a rare lung tumour

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ABSTRACT

Pulmonary blastomas are a rare aggressive neoplasm comprising 0.25-0.5% of all primary lung tumours. Morphologically they mimic foetal lung tissue before 4 months gestation. Study reports a case of 23-year-old male patient who came with heaviness in chest and breathing difficulty since one and a half months. On CT scan of chest, findings are suggestive of anterior mediastinal mass extending till right hila with cardiac displacement and compression. Histopathology confirmed the presence of pulmonary blastoma.

Keywords: Classic biphasic pulmonary blastoma, Lung tumour, Pulmonary blastoma

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INTRODUCTION

Pulmonary blastomas are a rare aggressive neoplasm comprising 0.25-0.5% of all primary lung tumours.¹ Morphologically they mimic foetal lung tissue before 4 months gestation.² First described by Barnard, they have since been divided into three subgroups: classic biphasic pulmonary blastoma (CBPB), well-differentiated foetal adenocarcinoma, also called monophasic pulmonary blastoma and pleuropulmonary blastoma of childhood.³ While well-differentiated foetal 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.

CASE REPORT

A 23-year-old male patient came to CTVS (cardio thoracic and vascular surgery) out-patient department with heaviness in chest and breathing difficulty since one

and a half months. There was history of occasional cough. There was no history of fever, night sweats, and wheeze or chest pain.

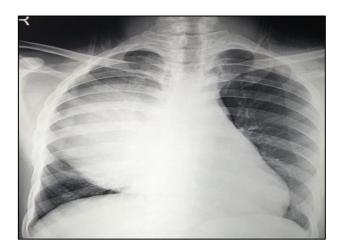


Figure 1: Chest X-ray of right-sided circumscribed mass occupying right middle zone and upper part of lower zone.

Also, there was no hoarseness of voice or hemoptysis. There were no associated co-morbidities and no history of any addictions. Family history was unremarkable. Baseline investigations of the patient were nonsignificant except for moderate anemia and mild thrombocytopenia. On chest X-ray a right-sided circumscribed mass was seen occupying right middle zone and upper part of lower zone with no evidence of any pleural effusion as shown in (Figure 1).



Figure 2: CT scan of thorax showing hypodense area measuring 153 x 106 mm in anterior mediastinal region extending till the right hila.

On CT (computed tomography) scan of thorax, hypodense area measuring 153 x 106 mm is seen in anterior mediastinal region extending till the right hila. Mass is abutting brachiocephalic veins, aorta, superior vena cava right pulmonary artery and cardiac chambers. Areas of necrosis are seen as shown in (Figure 2). 2D Echocardiography was normal. On examination, bilateral lungs were clear; trachea was central. There was no evidence of pleural effusion.

Patient was taken to operation theatre after clearance from the anaesthetist. Written informed consent was sought from the patient before operation. On exploration, a large anterior mediastinal mass was present extending from anterior mediastinum to the hilum of right lung, inferiorly extending to diaphragm and medially to pericardium, superior vena cava and innominate vein as depicted in (Figure 3).

Tumour was resected en mass with right lung and sent for histopathology examination as shown in (Figure 4). The biopsy report revealed ill-defined neoplastic growth showing biphasic pattern comprising epithelial and mesenchymal components consistent with pulmonary blastoma.



Figure 3: Intraoperative image of a large anterior mediastinal mass.



Figure 4: The mass after resection.

DISCUSSION

About 80% of CBPB occur in adults, and can occur at any age between birth and seventy years. CBPB shows a slight male preponderance and is common among smokers. Interestingly, our patient was a teenager, nonsmoker patient. Symptoms in CBPB are likely to seem like an upper respiratory infection, but recurrent, persistent symptoms require investigations. In CBPB, chest radiography is usually helpful and typically shows a well-demarcated peripheral or midlung mass with no definite lobar predominance. In this case also, a well-circumscribed lesion was seen in the right middle zone and upper part of lower zone which was confirmed on

chest CT scan showing an anterior mediastinal mass extending till the right hila with marked necrosis.

Immunohistochemistry plays a vital role in the diagnosis of CBPB. Each component can be clearly distinguished with each other when a combination of both epithelial and mesenchymal markers is used. In our case, complete excision biopsy was done, and a panel of immunohistochemical markers confirmed the biphasic pattern comprising epithelial and malignant mesenchymal components consistent with CBPB.

Surgical resection is the mainstay of treatment for the patients with PB. A number of cases of surgical approach with curative intent have been reported including an interesting case with a 24-year complete remission post-surgery.

Prognosis is poor with two thirds of patients dying within 2 years and only a 16%, 5-year survival. Prognosis is determined by size of the tumour at time of diagnosis, with tumours <5 cm doing better. Tumour metastasis and tumour recurrence despite resection both predict a poor prognosis. Unfortunately, 43% of tumours recur within 1 year with a propensity for sites such as brain and mediastinum.⁵ Recurrence tends to occur within 1 year after diagnosis or not at all.⁶

CONCLUSION

Study present a case of a classic biphasic pulmonary blastoma, a rare lung tumour, occurring at an earlier age and portending to poorer prognosis than other more common lung tumours. Recurrence after resection is high and regular surveillance is recommended especially within the first year. Our patient opted not to undergo adjuvant chemotherapy and remains disease free till last follow up.

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