

Case Report

Pulmonary blastoma: treatment through sleeve resection of the right upper lobe*

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ABSTRACT

Pulmonary blastoma is a rare lung tumor that is composed of malignant epithelial and mesenchymal cells. It presents a pattern of rapid growth. Herein, we report the case of a patient with hemoptysis and a mass in the right upper lobe. The patient presented limited pulmonary function, and fiberoptic bronchoscopy revealed invasion of the intermediate bronchus. The patient underwent sleeve resection of right upper lobe, a technique never before described. After 36 months of follow-up, the patient remained asymptomatic. We also review the literature regarding treatment, clinical aspects and pathology.

Keywords: Pulmonary blastoma/surgery; Pneumonectomy; Case reports [Publication type]

INTRODUCTION

Pulmonary blastoma is a rare lung tumor that presents a pattern of rapid growth and is associated with a poor prognosis. It is composed of malignant epithelial and mesenchymal cells that are morphologically similar to those of the embryonic lung.

CASE REPORT

A 52-year-old female patient, previously healthy, presented with mild hemoptysis. The patient denied any history of tuberculosis or smoking. Upon physical examination, the patient was pale, with no palpable lymph nodes, and chest auscultation

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revealed reduced breath sounds on the right side. Chest X-ray (Figure 1) revealed a juxtamediastinal right upper lobe mass with ill-defined borders. She was initially submitted to fiberoptic bronchoscopy, which revealed an endobronchial lesion located approximately 2 cm distal to the tracheal carina. This lesion obstructed the lumen of the right upper lobe bronchus and invaded the middle lobe bronchus, occluding approximately half of its lumen. No bleeding was detected during the examination. A biopsy and a bronchial lavage were performed. The results of mycological, mycobacterial and cytopathologic tests of the bronchial lavage were negative. The anatomopathological examination of the biopsy sample revealed a mesenchymal neoplasia. The patient was then submitted to a systemic staging, which revealed no alterations.

A computed tomography scan of the chest revealed atelectasis of the right upper lobe (caused by occlusion of the upper lobe bronchus) that extended up to the juxtacarinal portion of the main bronchus, accompanied by infiltration of the wall of the main left bronchus, which resulted in invasion of the right branch of the pulmonary artery and of the arch of the azygos vein. There was no sign of invasion of the chest wall or of pleural effusion (Figure 2). Tomographic findings led us to suspect invasion of the superior vena cava and azygos vein. Therefore, we performed a cavography, which did not reveal any involvement of these structures (Figure 3).

The evaluation of the expiratory reserve volume revealed forced vital capacity of 1.92 (62%) and forced expiratory volume in one second of 1.08 (40.8%). In the cardiologic evaluation, the patient was considered at low risk for the proposed procedure of surgical staging by cervical mediastinoscopy and sleeve resection of the right upper lobe.

A cervical mediastinoscopy was performed to evaluate the right, left and subcarinal paratracheal lymph node chains through an anatomopathological exam performed by freezing, the result of which was negative for neoplasia. The patient was then submitted to a right posterior lateral thoracotomy, entering the thoracic cavity through the fourth intercostal space. The inventory of the thoracic cavity showed an adherence between the upper lobe and the parietal pleura. In order to separate the two, an extrapleural dissection was performed. The

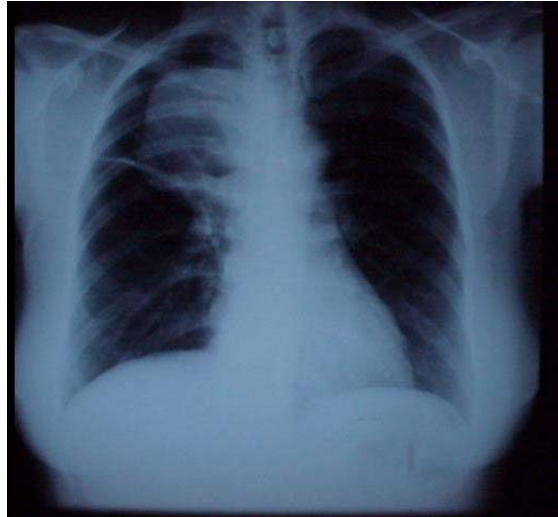


Figure 1 - Chest X-ray: mass in the right upper lobe in a juxtamediastinal position

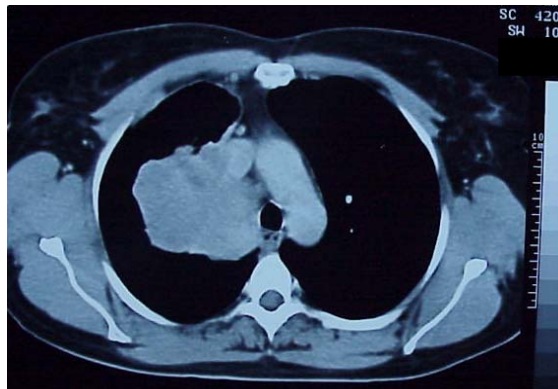


Figure 2 - Computed tomography: large mass in the right upper lobe and probable invasion of the superior vena cava

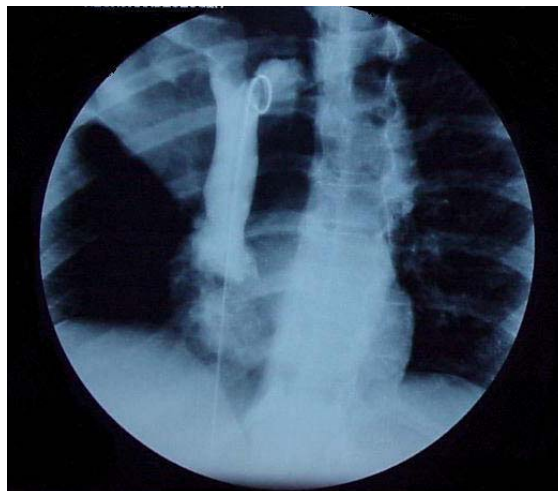


Figure 3 - Cavography: superior vena cava with regular walls suggesting absence of tumor invasion

interlobular portion of the pulmonary artery showed no signs of invasion. A sleeve resection of the right upper lobe was then performed using PDS 4.0 suture. The procedure lasted four hours, with 850 ml of bleeding. Analgesia was achieved through peridural nerve block with morphine.

The patient was discharged from the intensive care unit on postoperative day six and from the hospital on postoperative day nine. The anatomopathological and immunohistochemical analysis revealed a well-differentiated fetal lung adenocarcinoma, measuring 7.9 × 6.7 × 6.0 cm, with surgical borders and lymph nodes free of the neoplasia, which was staged as T2N0M0. The results of the tests for antibodies were as follows: positive for chromogranin; positive for synaptophysin; positive for somatostatin; positive for specific neuronal enolase; positive for keratin 7; positive for vimentin; positive for smooth muscle actin; negative for keratin 20; and negative for carcinoembryonic antigen.

DISCUSSION

There are two histological types of pulmonary blastoma: monophasic, or well-differentiated, fetal lung adenocarcinoma (composed only of epithelial cells); and biphasic pulmonary blastoma (composed of malignant mesenchymal tissue and epithelial cells). The monophasic type presents the better prognosis. Pulmonary blastoma occurs mainly in young women, most of whom present a peripheral mass (mean, 9 cm) with mediastinal involvement,⁽¹⁾ complaining of cough in 30% of cases and of hemoptysis in 20%. Recent studies suggest a correlation with smoking.⁽¹⁾ Since the first report by Barrett & Barnard, in 1945, approximately 200 cases of pulmonary blastoma have been reported and well documented in the literature.⁽¹⁾ We found no reports of primary sleeve resection in the literature. However, a case in which left sleeve pneumonectomy was used to treat stump recurrence after upper lobectomy was reported in the Japanese literature.⁽²⁾

Although it is an aggressive neoplasia usually diagnosed in advanced stages, surgical treatment is

mandatory,⁽³⁾ and chemotherapy is used in cases for which neoadjuvant therapy is recommended, as well as in nonsurgical cases or cases with involvement of surgical borders.⁽⁴⁾ In most health care facilities, radiotherapy is used to treat cases that do not respond to other forms of treatment.⁽⁵⁾ Some authors suggest that the combination of surgery, adjuvant radiotherapy and chemotherapy based on cisplatin and etoposide should be considered in the treatment of this neoplasia.⁽⁴⁾ Using an adjuvant protocol similar to the one used in the treatment of germ cell tumors (cisplatin, VP-16, uromitexan, ifosfamide and 64 Gy of mediastinal radiotherapy), a Swiss group reported a 33-month survival in a stage III-A patient (pT3N2M0).⁽¹⁾

Therefore, the treatment of the pulmonary blastoma should be submitted to a joint analysis from a pulmonologist, an oncologist and a thoracic surgeon for a better definition of the best treatment strategy. The present case illustrates an indication of treatment of pulmonary blastoma by lobectomy accompanied by sleeve resection based on spirometry, fibrobronchoscopic findings and surgical staging, without referral for adjuvant treatment. The patient remained asymptomatic after a 36-month follow-up period.

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