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Case Report

Primary pulmonary leiomyosarcoma: review of the literature and case report

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ABSTRACT

Primary pulmonary leiomyosarcoma (PPL) is an extremely rare malignant tumor. It has been revealed that PPL may originate from the smooth muscle of the pulmonary parenchyma, pulmonary arteries and bronchi. Patients with PPL may be asymptomatic or present with symptoms similar to those observed in other primary lung tumors. The present study reports the case of a 21-year-old woman who presented with orthopnea, medium effort dyspnea, so an extension study computed tomography (CT) is performed, finding a left thoracic tumor. Where a percutaneous biopsy is performed, concluding PPL, by image, it is considered unresecable due to pericardial infiltration, and it is sent to cycle of chemotherapy. Currently completed treatment with partial response to it, due to findings by image, ECOG and patient age, a case session is decided, and panpleuronectomy was performed.

Keywords: Primary pulmonary leiomyosarcoma, Sarcoma, Panpleuropneumonectomy

INTRODUCTION

Leiomyosarcomas are tumors of the smooth muscle cells that may originate in any location, but most often arise in the uterus, gastrointestinal tract and soft tissue.[1] PPL is an extremely rare malignant mesenchymal tumor that appears to originate from the smooth muscle cells of the bronchial and blood vessel wall.[2] The incidence has been reported to be less than 0.5% of all malignant pulmonary tumors and 30% of primary sarcomas of the lung.[3]

LPP presents with general symptoms of any other lung tumor, including hemoptysis, dyspnea, asthenia, chest and low back pain, and weight loss. However, because they are rare neoplasms, LPPs have been misdiagnosed with other diagnostic differences such as pulmonary emboli, or even cardiac neoplasms.[4]

Tumors can be treated by surgical resection, which is the primary and definitive mode of treatment. Surgical techniques consist of lobectomy, pneumonectomy, and bronchial sleeve resection, depending on the clinical stage. The role of other treatment methods has not yet been defined; however, radiochemotherapy is recommended in cases of incomplete resection with positive margins.[5]

Early detection and complete surgical resection of PPL have been demonstrated to significantly contribute to an increased survival time of patients with the disease.[6] We herein report a case of primary pulmonary leiomyosarcoma that was successfully resected by surgery after neoadjuvant chemotherapy.

CASE REPORT

A 21-year-old woman was admitted to the oncology hospital CMNSXXI (Mexico City, México) in April 2022 with a cough and the expectoration of white and yellow

sputum, which had been ongoing for 2 months, orthopnea, medium effort dyspnea.

The patient had been a smoker for 4 years, but had no family history of lung cancer. The vital signs of the patient were stable. Lung auscultation revealed decreased breath sounds on the left lung and fine crackling was heard at the base of the left lung. Other physical examinations were unremarkable.

Blood tests revealed that the patient's white blood count was 3.4×109 cells/l (normal range, 4.0-10.0×109 cells/l), albumin 4.3 gr/dl, Hemoglobin 13.4 gr/dl. Other laboratory examinations were all negative, including those for blood chemistry, blood tumor markers, and liver and renal function.

Enhanced computed tomography (CT) scans of the patient's chest revealed the presence of left lung with a voluminous lesion that occupies the entire lung, multilobulated, heterogeneous with a predominant hypodense density, linear calcifications of the septa with dimensions of 11.2 cm, compressing the main bronchus and segmental lower lobe. contralateral lung without injuries. mediastinum without nodal growths. mild pericardial effusion. left supraclavicular node of 9 mm. with a 31% decrease in dimensions compared to the previous study, partial response. mild pleural effusion. (Figure 1), by image, it is considered unresecable due to pericardial infiltration, and it is sent to cycle of chemotherapy.

Subsequently, the patient was administered with systemic chemotherapy, previously to resection, as follows: epirubicin, ifosfamide, and vincristine, suspended due to epirubicin allergy, starting second line with gemcitabine, docetaxel 6 cycles.

The following tomography was taken at the end of the cycles with a 31% decrease in dimensions compared to the previous study, partial response. mild pleural effusion. There were no metastatic lesions observed on abdominal CT and magnetic resonance imaging (MRI) of the brain.

Where a percutaneous biopsy is performed, demonstrated that the tumor was composed of spindle cells, with marked nuclear pleomorphism and numerous mitotic figures. Immunohistochemical staining demonstrated that the tumor expressed vimentin, smooth muscle actin, cluster of differentiation 34 (CD34) and actin, and did not express high and low molecular weight cytokeratins, epithelial membrane antigen, desmin and protein S-100. There was no epithelial differentiation observed in the tumor and the overall morphological features favored a high-grade sarcoma with evidence of smooth muscle differentiation, indicating a leiomyosarcoma. Thus, the final diagnosis was PPL.

We performed left posterolateral thoracotomy preservative of serratus, (Figure 2) and left panpleuropneumonectomy. A transoperative study of a pleural biopsy was performed with fibroconnective tissue with focal sclerosis, no neoplastic cells are observed in the examined material. Therefore, we decided to perform panpleuropneumonectomy (Figure 3) We found voluminous tumor depending on the left upper lobe which extends from the lateral thoracic wall where it invades visceral pleura, and extends towards the anterior mediastinum, infiltrating the main bronchus within 2 cm of the carina and pulmonary artery, firmly adhered to the pericardium without infiltrating it (Figure 4). The patient had an adequate evolution and it was decided to be discharged from the hospital a week after surgery.

The patient was still alive without recurrence at 3 months postoperatively (Figure 5).

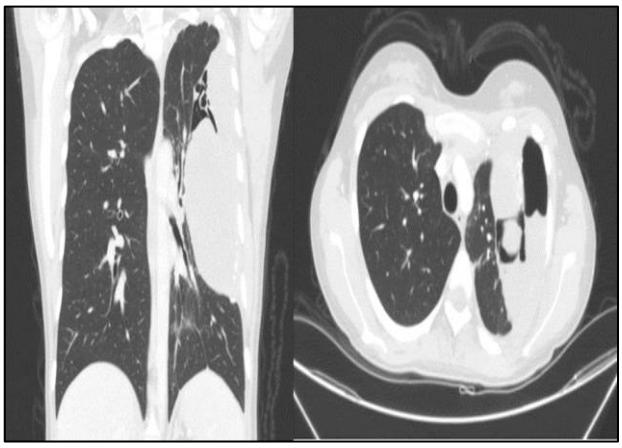

Figure 1: Enhanced computed tomography scans of left lung with a voluminous lesion that occupies the entire lung, multilobulated, heterogeneous with a predominant hypodense density, linear calcifications of the septa with dimensions of 11.2 cm, compressing the main bronchus and segmental lower lobe.

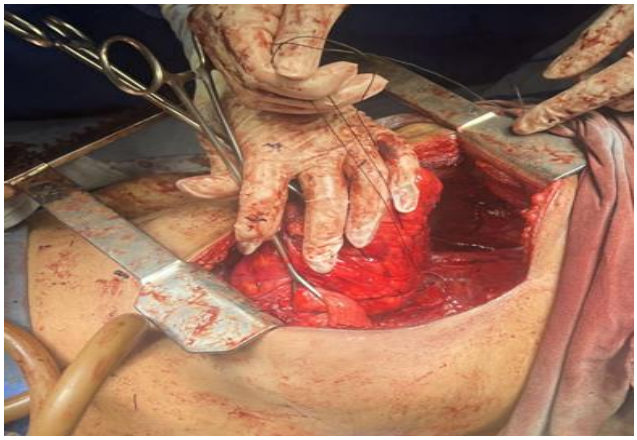

Figure 2: Left posterolateral thoracotomy preservative of serratus, left pulmonary artery ligation.

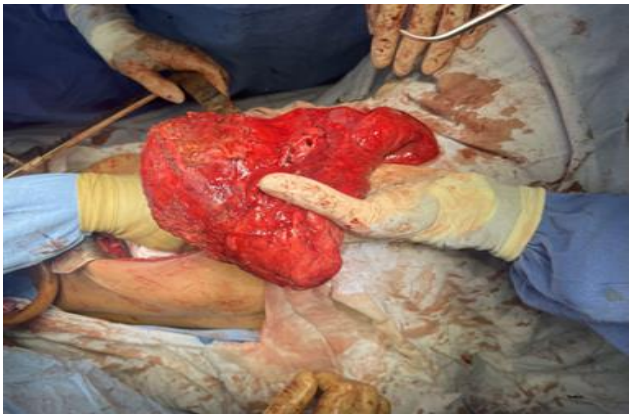

Figure 3: Lung being removed.

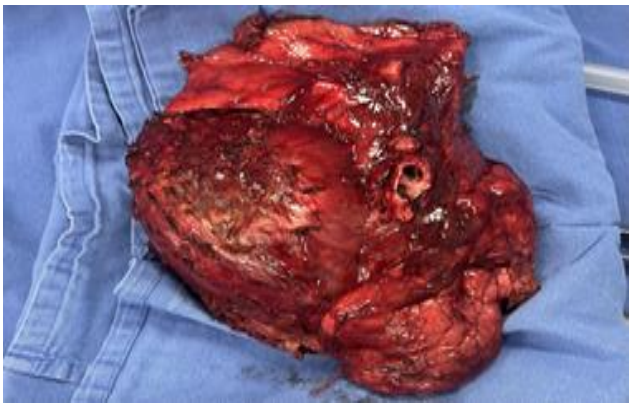

Figure 4: Product of left panpleuronectomy.

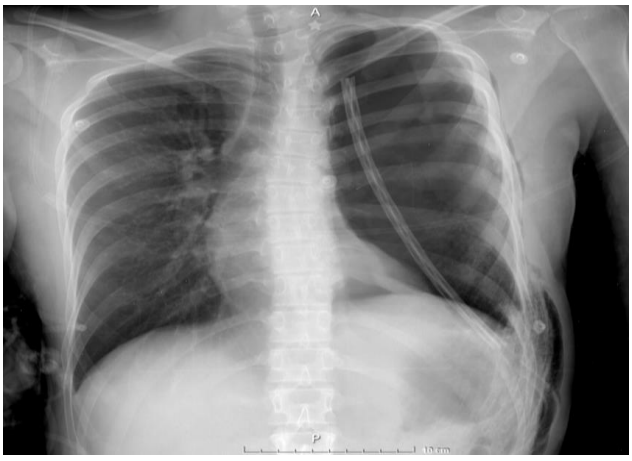

Figure 5: Chest x-ray after panpleuronectomy.

DISCUSSION

PPL is a mesenchymal tumor that has been observed to originate from the smooth muscle cells of the bronchial or blood vessel wall. PPL classified by type: Intraluminal, intrapulmonary and pulmonary vascular. The intrapulmonary type of PPL is the most common, and the pulmonary vascular type originates from the vascular wall and occurs in the pulmonary artery, where it may cause stenosis and obstruction. The majority of patients with PPL present with symptoms similar to those observed in other primary pulmonary tumors. In order to differentiate PPL from bronchogenic carcinoma, an excisional biopsy is required.[7]

Vimentin is present in the majority of mesenchymal cells and is a good mesenchymal tumor marker, which may additionally be used in the identification of sarcoma and carcinoma.

In the present case, immunohistochemical staining demonstrated that the tumor expressed vimentin, smooth muscle actin, cluster of differentiation 34 and actin, and did not express high and low molecular weight cytokeratins. Generally, leiomyosarcomas do not express carcinoembryonic antigen, cytokeratin, leukocyte common antigen, neuroendocrine filament and S100 protein.[8]

Treatment regimens for patients with PPL aim to achieve local and systemic control of the tumor, while preserving function and quality of life. If pre-operative staging demonstrates that there is no evidence of metastases, surgery is recommended, such as a lobectomy and pneumonectomy, which require resection of the chest wall, diaphragm or vascular structures. If an early complete resection is performed, the 5-year survival rate of patients is 50%, and there have been reports of patient survival 20 years post-resection.[9] Adjuvant radio or chemotherapy treatment is recommended in cases of incomplete resection, unresectable tumors and patients with increased histological malignancy, as in this case.

Prognostic indicators of PPL consist of tumor size, extent of bronchial invasion and degree of malignancy.

In the present study, malignant spindle cells were positive for S100 antigen, smooth muscle actin and vimentin, and were negative for cytokeratin 20, CD34, c-Kit, desmin, epithelial membrane antigen, myogenin. Therefore, the results of the present study are consistent with the existing literature.[10]

In some cases, the diagnosis of sarcoma is considered either too late, or not at all. The definitive PPL diagnosis requires a biopsy followed by histopathologic examination with appropriate immunohistochemical analyses. Additionally, the early detection, diagnosis, and complete surgical resection of PPLs contribute significantly to increased patient survival times[11]. For this reason, clinicians should be aware of these rare lesions, especially when the symptoms of thromboembolic disease fail to resolve with anticoagulation therapy.

CONCLUSION

In conclusion, PPL is a rare tumor that grows rapidly. It may be challenging to differentiate PPL from other pulmonary tumors due to the lack of specific manifestations. A pre-operative diagnosis of PPL is

considered following the results of a sputum smear, lung biopsy or bronchoscopic examination; however, diagnosing PPL from these methods may also be challenging. In order to differentiate a primary pulmonary leiomyosarcoma from bronchogenic carcinoma, an excisional biopsy is required.

The present case emphasizes the important role of pathological and immunohistochemical results in differentiating between PPL and bronchogenic carcinoma. The goal of treatment is to obtain local and systemic control of the sarcoma, while preserving functioning and quality of life. If preoperative evaluation reveals no evidence of metastases, then treatment is surgical. However, if preoperative evaluation reveals evidence of metastasis, the radiation therapy, chemotherapy or a combination of the two is required, like in this case. An increased awareness of PPL leading to an early diagnosis and the performance of a complete surgical resection with adjuvant radio and chemotherapy in selected patients may improve the prognosis of patients with PPL.

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