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clinical practice

Resection of a giant mediastinal leiomyosarcoma

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SUMMARY: **Resection of a giant mediastinal leiomyosarcoma.**

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Primary leiomyosarcomas of the lung are tumors. We report a case of 49-year old female with history of cough, breathless at rest, right sided chest pain. Chest CT showed a huge (16 cm) mediastinal mass located on the right mediastinum encasing the right main pulmonary artery and infiltrating the main right bronchus and pericardium. The tumor was resected with combined pericardiectomy and pnemonectomy via hemiclamshell incision. This surgical access provided an adequate exposure of the chest "blind zones" and it allowed a radical and safe surgical resection of lung, pleura, pericardium and diaphragm. The final diagnosis showed a low grade differentiation leiomyosarcoma.

KEY WORDS: Giant leiomyosarcoma - Lung leiomyosarcoma - Mediastinal mass - Pericardiectomy - Pnemonectomy - Hemiclamshell incision.

Introduction

Primary leiomyosarcomas of the lung are extremely rare tumors arising from the smooth muscles of the pulmonary interstitium, bronchi and blood vessels (1). The majority of the leiomyosarcomas arises from the peribronchial smooth muscle cells present around the hilum and most frequently from the larger bronchi of the left lower lobe (2). Clinical presentation may vary with symptoms seen in other primary lung neoplasms or they may be asymptomatic and found incidentally on imaging studies. Herein, we report a case of resection of a giant mediastinal leiomyosarcoma located on right mediastinum infiltranting the lower pulmunary lobe, the main right bronchus and the pericardium, compressing the pulmonary artery and veins.

Case report

A 49-year-old female nonsmoker presented with a 2-month history of dry cough, breathless at rest, right sided chest pain and bilateral leg edema. She denied any history of fever, hemoptysis, and loss of weight or other extrapulmonary symptoms. She also denied any history of pulmonary tuberculosis, asthma or chronic obstructive pulmonary disease. She received hysterectomy for benign leiomyoma several years before, she was normotensive and nondiabetic. General physical examination was unremarkable besides bilateral leg edema and nail clubbing; no pallor, icterus, cyanosis, or lymphadenopathy. Examination of the respiratory system revealed decreased breath sound in right hemithorax. Other systemic examination was unremarkable. Biochemical investigation and hematological parameters were within normal limits. Imaging studies of abdominal and

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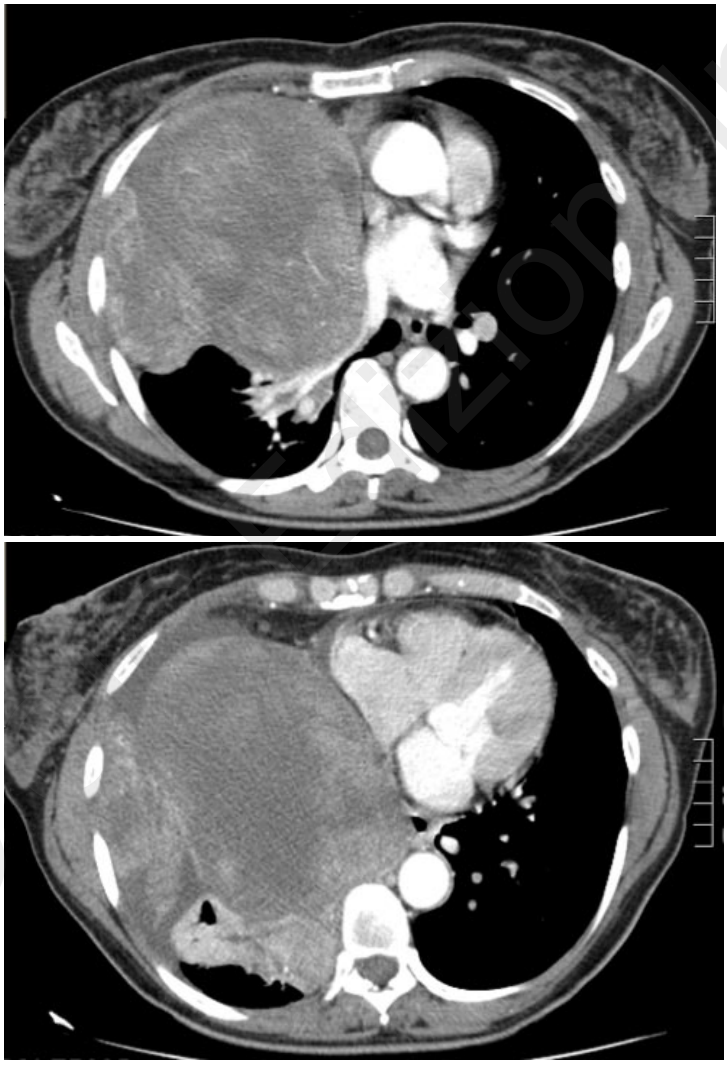
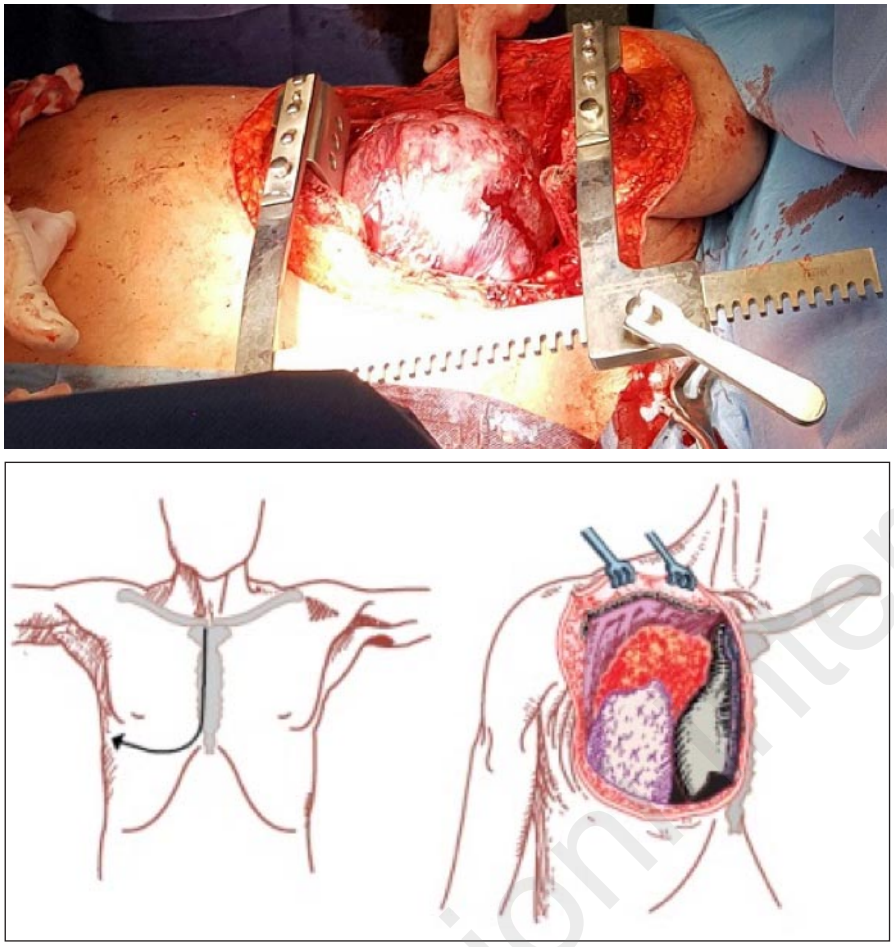
pelvic organs were essentially within normal limit. Chest computed tomography showed a huge mediastinal mass with the largest diameter of 16 cm located on the right mediastinum encasing and causing narrowing of the right main pulmonary artery (Figures 1, 2). The CT scan showed also 2 solid lesion on the left lung measuring 2 cm and 1,5 cm. These two lesions were positive also at PET-CT. The mass is also extending distally around the right main bronchus. Fiberoptic bronchoscopy revealed mucosal infiltration and almost complete obstruction of the right main bronchus. The mass was diagnosed as a leiomyosarcoma by CT-guided biopsy. Surgical resection was considered the more appropriate treatment option as palliative treatment mainly to prevent the more severe involvement of the heart. After special multidisciplinary discussion the operability of the tumor and the surgical strategy were assessed. Eventually, we agreed that exploratory surgery for resection of the large mass using cardiopulmonary bypass should be performed. The tumor was so huge that it pushed the heart and mediastinum to the right side (3). Surgical access to the mass was first chose as left anterolateral thoracotomy through the 4th intercostal space in the semi-lateral decubitus position, which was extended to hemiclamshell approach since the upper pole of tumor was difficult to expose and in order to obtain a better exposition of the mass, of the pericardium and the diaphragm (Figures 3, 4). The tumor was fused to the lungs, requiring a partial resection of the right middle and lower lobes. No adhesions between the



Figures 1, 2 - Chest CT shows a mass measuring 16 × 14 cm in diameter, in contact with the rigth lung, mediastinal pleura, and parietal pleural. Encasing and causing narrowing of the right main pulmonary artery.



Figures 3, 4 - Hemiclamshell incision gives a better exposition of the mass, of the pericardium and the diaphragm.

tumor and the diaphragm were found. The right phrenic nerve was invaded by the tumor and it was resected. The tumor moved the heart on the right side and widely infiltrated the pericardium and the right main bronchus so a large right pericardial resection was performed and a bovine pericardial prosthesis was placed (4). The excised specimen shows a tumor measuring 16 × 14 × 15 cm and the weight of the tumor was 1,535 g (Figure 5). At last extra-pericardial pneumonectomy was done.

Discussion

Primary pulmonary leiomyosarcoma is rare and is the most common pathological type of sarcoma in the lungs. They are mostly found in adults, with a male to female ratio of 1,5:1 (5). The bilateral pulmonary lesion usually suggests metastatic malignancy.

In our case the tumor was located in the anterior-inferior mediastinum and protruded into pleural cavity. It needs to be distinguished with solitary fibrous tumors, tumors of pleural origin, chest wall tumors, and metastatic tumors. The only way to definitive diagnosis was pathological diagnosis obtained by CT guided biopsy (6, 7). It has been suggested that pulmonary leiomyosarcoma in female patients should never be considered primary neoplasm as there are reports of PPL arising in patients with history of hysterectomy for benign leiomyoma several years before presentation. In this case the hystopathologic feature cannot allow to assess the origin of the tumor due to the low grade cell differentiation (8, 9).

Surgical resection is the mainstay of therapy for pulmonary leiomyosarcoma and is usually curative for small and well-differentiated sarcoma. Metastatic stage, unresectability, tumor diameter of >10 cm, and grade 3 diseases were predictable of poor survival (10, 11).

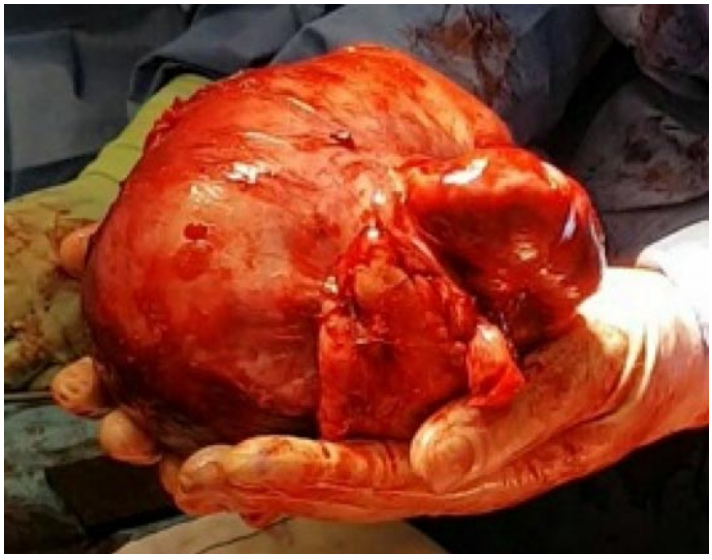


Figure 5 - Gross pathologic finding of the tumor: an encapsulated mass measuring 16 × 14 × 15 cm.

Conclusion

Although anterolateral thoracotomy is the standard approach for this type of lesions, hemiclamshell approach gives a better access to upper thoracic cavity compared with anterolateral thoracotomy. In our case, we took anterolateral thoracotomy as first choice and extended the incision to hemiclamshell approach when the upper pole of tumor was difficult to expose. As already highlighted unusual surgical access can provides an adequate exposure of the chest "blind zones" and allows a radical and safe surgical resection of lung, pleura, pericardium and diaphragm (12). In our case the rationale of surgery was not therapeutic due to bilateral lesions but surgery was palliative mainly to prevent severe and acute heart impairment (13).

Disclosure

The Authors report no conflicts of interest in this work. The informed consent was obtained to this case report.

Acknowledgments and credits

None to declare.

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