A Rare Case of Leiomyosarcoma of Lung

Karishma Vahora, Naresh Patel**, Tushar Patel***, Rushi Patel***, Yogin Mistry*****

Abstract

Leiomyosarcoma is a rare type of malignant soft tissue sarcoma derived from smooth muscle cells. A 60 year old male patient, chronic smoker, presented with dull right sided chest pain and associated with anterior chest wall swelling since 20 days. A CECT thorax was done with high risk consent which showed 56x62x61 mm soft tissue density lesion with speculated margin involving apical and posterior segment of right upper lobe. The lesion showed cut off of right upper lobe bronchus. Similar lesions were noted in apico-posterior and anterior segment of right upper lobe. He underwent bronchoscopy with biopsy. Biopsy revealed high grade sarcoma of lung lesion. IHC was done for CK, Vimentin, Desmin, S-100, Actin of which Vimention and Actin were positive hence proving the diagnosis of Leiomyosarcoma of lung.

Key words: Bronchoscopy, CECT, Leiomyosarcoma

Introduction:

Leiomyosarcoma is a rare type of malignant soft tissue sarcoma derived from smooth muscle cells. Smooth muscle is present in the bronchi and blood vessels of the lung but it is generally believed that most leiomyosarcomas arise from the bronchi. Further, they most frequently arise from the larger bronchi where most smooth muscle is present.[1] These tumors are most common in uterus, stomach, small intestine and retroperitoneum. Sarcomas are malignant tumors arising from mesenchymal cell lines. Of all soft tissue sarcomas approximately 5-10 percent are leiomyosarcomas.[2] Leiomyosarcomas are classically been subdivided into 3 groups for prognostic and treatment purposes: leiomyosarcoma of somatic soft tissue; cutaneous leiomyosarcomas and leiomyosarcoma of vascular origin.[3] The prognosis is poor, with survival rates among the lowest of all soft tissue sarcomas.[4] Leiomyosarcoma resembles bronchial carcinoma in its clinical presentation and radiological appearance, and differentiation from carcinoma is possible only on histological grounds.[5]

* Resident
** Professor & Head
*** Associate Professor
**** Senior Resident, Department of Respiratory Medicine, GCS Medical College, Hospital & Research Centre, Ahmedabad, Gujarat, India
Correspondence : Dr. Karishma Vahora
E-mail : karishmavhora@yahoo.in

Case report:

A 60 year old male patient, who had smoked 20 bidies per day for 40 years, presented with dull right sided chest pain and associated with anterior chest wall swelling since 20 days. On examination; he was vitally stable and afebrile. The swelling of size of a fist, non-tender and temperature was normal by palpatory method. The swelling was fixed to anterior chest wall. There was no clubbing of finger or cyanosis. Systemic examination revealed mild hepatomegaly and auscultation revealed bilateral air entry present and clear. Chest x-ray showed shadow in right lung field (fig-1). Routine investigation revealed raised leucocyte count (32010 / cmm) and raised creatinine (1.91 mg / dl). Rest blood investigations were normal. USG of local part showed echogenic lobulated lesion of size 60x40 mm with internal vascularization. A CECT thorax was done with high risk consent which showed 56x62x61 mm soft tissue density lesion with speculated margin involving apical and posterior segment of right upper lobe. The lesion showed cut off of right upper lobe bronchus. Similar lesions were noted in apico-posterior and anterior segment of right upper lobe; largest measuring 20x20 mm. Mediastinal

lymph nodes were also found to be enlarged. There was a lobulated heterogeneously enhancing soft tissue density lesion of size 66x82 mm on anterior chest wall involving body of sternum. Right sided minimal pleural effusion of max thickness 12 mm was noted (fig-2, 3,). He was primarily treated with higher antibiotics and I.V. fluids. Sputum AFB under RNTCP and culture were negative. He underwent bronchoscopy with biopsy. Biopsy revealed high grade sarcoma of lung lesion. IHC was done for CK, Vimentin, Desmin, S-100, Actin of which Vimention and Actin were positive hence proving the diagnosis of Leiomyosarcoma of lung. Patient was referred to oncologist who advised chemotherapy (Doxorubicin and fosfomide). Unfortunately patient could not receive any chemotherapy and succumbed to death within few months.

Discussion:

Present case report is rare as the leimyosarcoma of lung is not a common entity. Primary pulmonary leiomyosarcoma is an extremely rare malignant mesenchymal tumor that appears to originate from the smooth muscle cells of the bronchial and blood vessel wall. Pulmonary leiomyosarcoma accounts for <0.5% of all malignant pulmonary tumors. Surgical strategies consist of lobectomy, pneumonectomy and bronchial sleeve resection. The role of other treatment methods has yet to be defined; however, radiochemotherapy is recommended in cases of incomplete resection and malignancy. 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.

Cytokeratin is an epithelial marker. Therefore, vimentin and cytokeratin may be used together to

Figure1: X-ray chest (PA view)

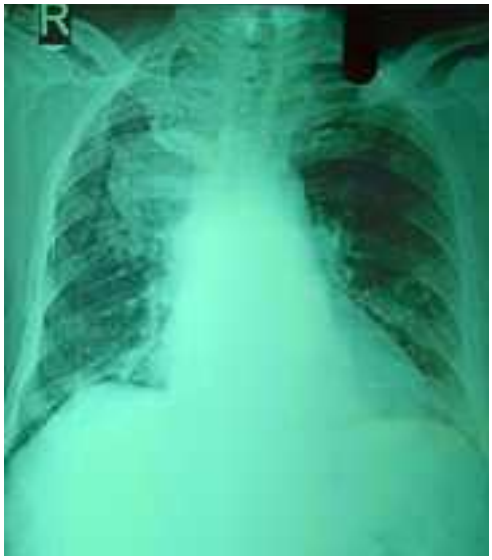

Figure 2: CECT Thorax

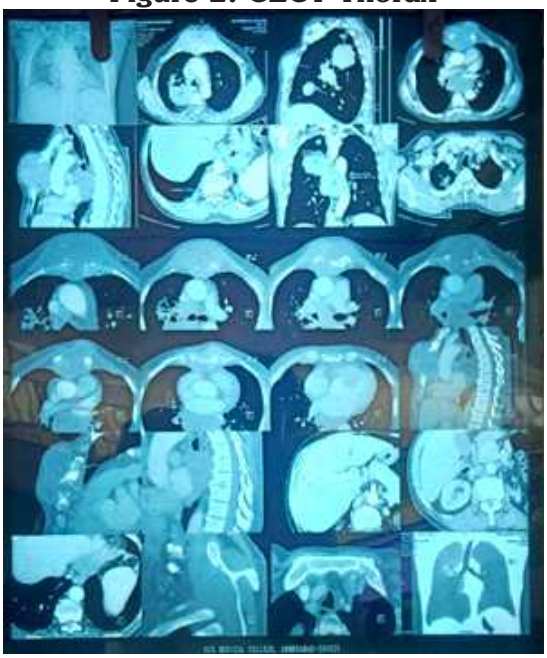

Figure 3: CECT Thorax

distinguish between the majority of epithelial and mesenchymal tumors. 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. A diagnosis of pulmonary leiomyosarcoma should only be considered if signs of an occult tumor are not noted in any other bodily location. In women, it is particularly important to observe if there is a tumor present in the uterus. Following a tissue biopsy that confirms pulmonary leiomyosarcoma, pre- operative staging of the tumor may be considered. Generally, this consists of CT of the lungs and MRI of the primary lesion to determine if the tumor has metastasized, which is uncommon in leiomyosarcomas and is typically observed in the advanced stages of the disease.[6] 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.

Conclusion:

Primary pulmonary leiomyosarcoma is an extremely rare malignant mesenchymal tumor. An increased awareness 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 pulmonary leiomyosarcoma.

References:

1. Glennie, J. S., Harvey, P., and Jewsbury, P. (1959). Two cases of leiomyosarcoma of the lung. Thorax, 14, 327.
2. Gustafson P, Willen H, Baldetrop B, et al. Soft tissue leiomyosarcoma: a population-based epidemiologic and prognostic study of 48 patients, including cellular DNA content. Cancer 70:114, 1992.
3. Weiss SW, Goldblum JR. enzinger and Weiss's soft tissue tumors fourth edition. Philadelphia, Mosby harcort, 2001.
4. Mankin, HJ, Casas-Ganem, J, Kim, JI, et al. Leiomyosarcoma of somatic soft tissues. Clin Orthop Relat Res. 2004 Apr;(421):225-31.
5. Dyson, B. C. and Trentalance, A. E. (1964). Resection of primary pulmonary sarcoma. Journal of Thoracic and Cardiovascular Surgery, 47, 577.
6. Ramanathan T. Primary leiomyosarcoma of the lung. Thorax. 1974;29:482–489. doi: 10.1136/thx.29.4.482.