Case Report
Published: 25 Mar, 2019

Journal of Clinical Case Reports

Primary Pulmonary Leiomyosarcoma Resection with Five-Year Relapse-Free Survival – Case Report

*Momozane T[1,2], Fukui E[1], Funaki S[1] and Shintani Y[1]**

[1]Department of General Thoracic Surgery, Osaka University, Japan

[2]Department of Thoracic Surgery, Yao Municipal Hospital, Japan

Abstract

A primary pulmonary sarcoma is relatively rare and the clinicopathological features are not well understood as compared with pulmonary carcinoma. Here, we report a non-elderly patient with a primary pulmonary leiomyosarcoma who underwent resection with a relapse-free survival for more than 5 years, along with a review of related literature. A 32-year-old male was found to have a well-circumscribed nodular shadow in the left lung field on a chest roentgenogram at medical screening. Chest computed tomography was not useful for differential diagnosis, while marked accumulation of 18F-fluorodeoxyglucose in the tumor was observed in positron emission tomography findings, indicating malignancy. A left upper lobectomy was performed and the post-operative pathological diagnosis was pulmonary leiomyosarcoma, T2aN0M0, stage IB. No post-operative treatment was given and the patient was followed for over 5 years, during which there was no evidence of recurrence or metastasis. A rapid growing well-defined nodule with abnormal FDG uptake in a non-elderly patient should be suspected as a primary pulmonary sarcoma and considered for resection.

Keywords: Lung; Young Patients; Sarcoma; Leiomyosarcoma; FDG-PET

Introduction

A primary pulmonary sarcoma is a relatively rare tumor [1,2]. The annual report of The Japanese Association for Thoracic Surgery in 2014 noted that the number of operations for primary pulmonary malignancy was 38,444, of which 40 were for sarcoma, a frequency of approximately 0.1% [3]. Thus, the clinicopathological features are not well understood. Herein, we report an adult patient who underwent resection of a primary pulmonary leiomyosarcoma with relapse-free survival for more than 5 years, along with a review of related literature.

Case Presentation

A small nodular shadow was found in the left lower lung field of a 32-year-old male in chest roentgenogram images obtained at a medical screening examination (Figure 1a). No such finding was noted in images obtained 1 year prior. Computed tomography (CT) imaging of the chest was performed, which showed a well-defined and smooth nodule measuring 21x20 mm in the inferior lingular segment of the left lung (S5), suspected to be a hamartoma, pneumo-cytoma, small cell lung carcinoma, or other similar type of mass (Figure 1b). CT findings did not show indications of malignant or benign status, and the patient reported no symptoms. Furthermore, there was no significant medical history and the patient had never smoked. A physical examination showed no remarkable findings, while laboratory results, including peripheral blood count, serum chemistry, and serum tumor markers, including carcinoembryonic antigen, cytokeratin 19 fragment, and pro-gastrin-releasing peptide, were all normal. An FDG-PET examination showed abnormal uptake in the nodule with a maximum standardized uptake value (SUV max) of 5.4 (Figure 1c), which was not observed at other sites. Based on the high SUV max in the nodule shown by FDG-PET, we suspected a malignant tumor. Bronchoscopy was not performed. A left lingular segmentectomy was promptly performed for diagnosis, which proceeded to a left upper lobectomy procedure with mediastinal lymph node sampling under video-assisted thoracoscopy, since intraoperative frozen section analysis findings led to a diagnosis of malignant tumor, suspected to be a sarcoma. The operative time was 200 minutes and blood loss was 10mL.

A pathological examination of the surgical specimen showed a white colored well-defined mass sized 20x13x33-mm (Figure 2a). Microscopically, the tumor consisted of spindle cells with irregular hyperchromatic nuclei (Figure 2b and 2c). Nuclear atypia and mitosis were few, and no necrosis was

OPEN ACCESS

*Correspondence:
Shintani Y, Department of General Thoracic Surgery, Osaka University Graduate School of Medicine, 2-2 (L5) Yamadaoka, Suita City, Osaka, 565-0871, Japan.
Tel: +81-6-6879-3164
Fax: +81-72-924-4820
E-mail: yshintani@thoracic.med.osaka-u.ac.jp

Received Date: 13 Feb 2019
Accepted Date: 21 Mar 2019
Published Date: 25 Mar 2019

Citation: Momozane T, Fukui E, Funaki S, Shintani Y. Primary Pulmonary Leiomyosarcoma Resection with Five-Year Relapse-Free Survival – Case Report. J Clin Case Rep. 2019; 2(1): 1019.

ISSN 2643-8194

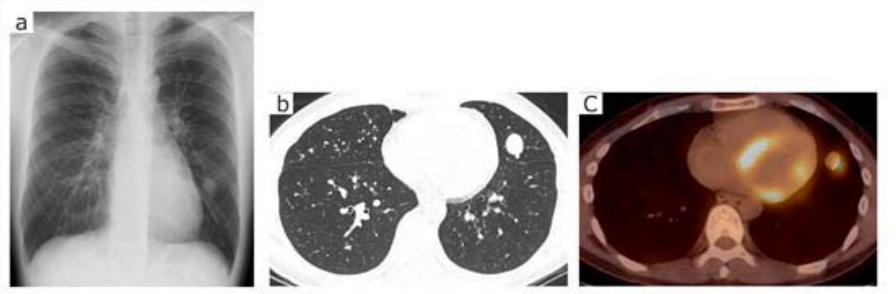

Figure 1: Images obtained prior to surgery. a) Chest radiograph showing nodule shadow in left lower field; b) Chest computed tomography showing 21×20 mm nodule in inferior lingular segment of left lung (S5); c) PET scan revealing marked accumulation of FDG in nodule with maximum standardized uptake value of 5.4.

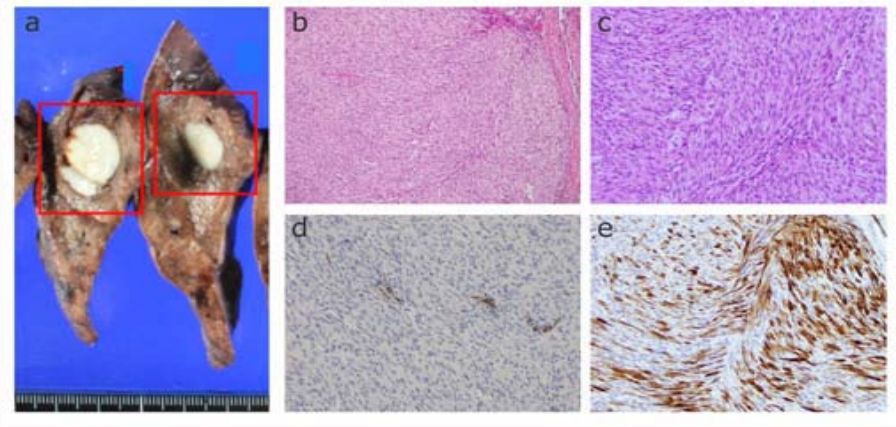

Figure 2: a) Macroscopic findings. Resected specimen showing a well-defined 20×13×33 mm solid tumor in inferior lingular segment of left lung (S5); b) Histological findings. Hematoxylin-eosin staining indicating that tumor consists of spindle tumor cells (×100); c) Magnification of part shown in (a) (×200); d) Positive immunohistochemical staining for SMA (×200); e) Positive immunohistochemical staining for desmin (×200).

seen. Immunohistochemistry results were positive for smooth muscle actin (SMA) and desmin (Figure 2d and 2e), while calponin, CD34, S-100, D2-40, and CK-AE1/AE3 were negative. Ki-67 was positive in only approximately 3-5% of the tumor cells. The pathological diagnosis was well-differentiated leiomyosarcoma, histological low grade. No lymph node metastasis was noted. The pathological stage was confirmed to be pT2aN0M0PL0D0E0PLC(-)PM0, stage IB in accordance with the TNM classification, 7th edition, from the Union for International Cancer Control. The postoperative course was uneventful and the patient was discharged on postoperative day 11. Thereafter, he was followed with no adjuvant therapy and was alive without recurrence for more than 5 years, with CT and FDG-PET screenings continued after the operation.

Discussion

A sarcoma is derived from cells of mesenchymal origin, such as bone, cartilage, fat, muscle, blood vessels, and others, with the majority found in soft tissue (nearly 75%), while 10% are related to bone [4]. There are more than 50 different histopathological subtypes of soft tissue sarcomas, including leiomyosarcoma [5]. A soft tissue sarcoma shows dissemination by a hematogenous route, with metastasis frequently found in the lungs and approximately 10% of affected patients showing lung metastasis at the time of diagnosis [6]. Therefore, careful clinical history taking and a total body survey are necessary to exclude the possibility of a primary tumor elsewhere in the body of a patient with a pulmonary sarcoma. In the present case, no new lesions appeared in other parts of the body more than 5 years after the initial resection, thus the tumor was considered to be a primary sarcoma of the lung.

Cases of primary pulmonary sarcoma have been reported to comprise less than 1% of lung malignancy cases [1,2], with a more recent report also noting the same frequency [3]. Although Spraker et al. presented a large-scale cohort study in 2013 [7], due to low frequency, other previous reports showed findings of approximately 20 cases each [8-14]. Selected prior studies that investigated primary pulmonary sarcoma occurrence are shown in Table 1. In those, the mean age at onset was approximately 50 years old, though with a wide range. Furthermore, more than 50% of the affected patients were symptomatic, such as cough, shortness of breath, hemoptysis, and chest pain, while the tumor diameter was also large. Accurate diagnosis of a primary pulmonary sarcoma is not easy without thoracotomy findings and a correct preoperative diagnosis was noted in 10-40% of those reported cases. The first choice of treatment is typically surgery, similar to primary lung carcinoma and soft tissue sarcoma, with the resection protocol including lobectomy, bilobectomy, and pneumonectomy with or without extension to peripheral organs, such as the chest wall, diaphragm, pericardium, phrenic nerve, and left atrium, and complete resection was achieved in 50-90% of those cases. At least one positive lymph node was noted in 5-30% of the reported patients and distant metastasis was uncommon. The majority of the resected tumors were histologically classified as leiomyosarcoma, such as in the present case, malignant fibrous histocytoma, or fibrosarcoma, and the histologic subtype did not have an influence on survival. Malignancy grade, shown in all cases of soft tissue sarcoma, based on differentiation, necrosis, and

Table 1: Summary of the selected previous reports investigating primary pulmonary sarcoma.

Author (Year)	N	Period	Mean or Median Age at Diagnosis (Yr, range)	Preoperative Diagnosis Rate (%)	Mean or Median Tumor Size (cm, range)	Lymph Node Metastases (%)	Distant Metastases (%)	Completed Resection (%)	5-Year Survival (%)	Prognostic Factor
Janssen (1994)	22	1959-1991	49.5 (4-76)	9.1	4 (1.5-20)	NA	9.1	50	44	Tumor size Tumor grade
Attanoos (1996)	14	1966-1995	52 (20-74)	NA	12 (2-25)	NA	21.4	NA	NA	NA
Bacha (1999)	23	1981-1996	51 (20-78)	30.4	5.2 (0.9-12.0)	30.4	NA	60.9	69 (resected)	Complete resection
Regnard (1999)	24	1973-1997	48 (14-73)	37.5	9 (4-18)	25 (resected)	NA	83.3	42	Complete resection stage
Keel (1999)	26	NA	48 (18-75)	NA	NA	15.4	7.7	NA	NA	NA
Porte (2000)	18	1982-1998	50 (19-73)	44.4	8.05 (2.5-15)	5.6	0	88.9	43	Stage
Etiennne-Mastroianni (2002)	12	1970-1999	53 (16-93)	25	6 (2.5-16)	NA	0	NA	38	NA
Spraker (2013)	365	1988-2008	63 (20-85)	NA	(> 5cm; 57%)	16	NA	NA	35	Patient age tumor size tumor grade treatment type

NA: Not Avaliable; Resected: Percentage only in resected cases.

mitotic rate [4], was high in most cases. The overall 5-year survival rate ranged from 30-40%, though varied greatly depending on clinicopathological features. Cases of incomplete resection and distant metastasis had poor prognosis, with resectability, tumor diameter, tumor grade, and stage considered to be prognostic factors. Some of those previous reports also suggested that a primary pulmonary sarcoma is a locally advanced but resectable tumor, and that early diagnosis and treatment are important for survival.

Since most of the previous reports were published around 2000, those findings may not fully reflect the current situation. Asymptomatic and small-sized primary pulmonary sarcoma cases might be more often diagnosed recently because of increased use of medical screening [15]. On the other hand, primary pulmonary sarcoma tumors in previous reports were often found to be locally advanced and this type of tumor may show rapid local progression. In the present case, a nodular shadow was clearly shown in chest roentgenogram findings within the course of 1 year. Although we found that chest CT was not useful for differential diagnosis because of a lack of specific manifestations, FDG-PET revealed abnormal uptake in the nodule, indicating malignancy. In the field of orthopedic medicine, a soft tissue sarcoma can be diagnosed conventionally using either CT or magnetic resonance imaging (MRI) results, while more recently PET-CT has been shown to have high sensitivity and specificity for early diagnosis, as well as staging and monitoring of potential malignancy [16–18]. We performed a thoracotomy, which allowed prompt diagnosis of a malignant tumor, suspected to be a sarcoma, and the mass was completely resected using a lobectomy approach. The pathological diagnosis of the resected specimen was a well-differentiated histologically low-grade leiomyosarcoma and the patient was negative for lymph node metastasis. A good prognosis was expected and the patient was alive without recurrence more than 5 years after the operation.

Conclusion

Primary pulmonary sarcomas have potential to show rapid local progression, while complete resection provides a good prognosis. A rapid growing well-defined nodule with abnormal FDG uptake in a non-elderly patient should be suspected as a primary pulmonary sarcoma and considered for resection.

References

1. Guccion JG, Rosen SH. Bronchopulmonary leiomyosarcoma and fibrosarcoma. A study of 32 cases and review of the literature. Cancer. 1972; 30: 836–847.
2. Cameron EW. Primary sarcoma of the lung. Thorax. 1975; 30: 516–520.
3. Committee for Scientific Affairs TJA for TS. Thoracic and cardiovascular surgery in Japan during 2014 : Annual report by The Japanese Association for Thoracic Surgery. Gen Thorac Cardiovasc Surg. 2016; 64: 665–697.
4. Casali PG, Abecassis N, Bauer S, Biagini R, Bielack S, et al. Soft tissue and visceral sarcomas: ESMO-EURACAN Clinical Practice Guidelines for diagnosis, treatment and follow-up. Ann Oncol. 2018; 29: iv51-iv67.
5. López-Pousa A, Martin Broto J, Martinez Trufero J, Sevilla I, Valverde C, et al. SEOM Clinical Guideline of management of soft-tissue sarcoma (2016). Clin Transl Oncol. 2016; 18: 1213–1220.
6. Christie-Large M, James SLJ, Tiessen L, Davies AM, Grimer RJ. Imaging strategy for detecting lung metastases at presentation in patients with soft tissue sarcomas. Eur J Cancer. 2008; 44: 1841–1845.
7. Spraker MB, Bair E, Bair R, Connell PP, Mahmood U, et al. An analysis of patient characteristics and clinical outcomes in primary pulmonary sarcoma. J Thorac Oncol. 2013; 8: 147–151.
8. Janssen JP, Mulder JJ, Wagenaar SS, Elbers HR, van den Bosch JM. Primary sarcoma of the lung: a clinical study with long-term follow-up. Ann Thorac Surg. 1994; 58: 1151–1155.
9. Attanoos RL, Appleton MA, Gibbs AR. Primary sarcomas of the lung: a clinicopathological and immunohistochemical study of 14 cases. Histopathology. 1996; 29: 29–36.
10. Bacha EA, Wright CD, Grillo HC, Wain JC, Moncure A, et al. Surgical treatment of primary pulmonary sarcomas. Eur J Cardiothorac Surg. 1999; 15: 456–460.
11. Régnard JF, Icard P, Guibert L, de Montpreville VT, Magdeleinat P, et al. Prognostic factors and results after surgical treatment of primary sarcomas of the lung. Ann Thorac Surg. 1999; 68: 227–231.
12. Keel SB, Bacha E, Mark EJ, Nielsen GP, Rosenberg AE. Primary pulmonary sarcoma: a clinicopathologic study of 26 cases. Mod Pathol. 1999; 12: 1124–1131.
13. Porte HL, Metois DG, Leroy X, Conti M, Gosselin B, et al. Surgical treatment of primary sarcoma of the lung. Eur J Cardiothorac Surg. 2000; 18: 136–142.

14. Etienne-Mastroianni B, Falchero L, Chalabreysse L, Loire R, Ranchère D, et al. Primary sarcomas of the lung: a clinicopathologic study of 12 cases. Lung Cancer. 2002; 38: 283–289.

15. Wood DE, Kazerooni EA, Baum SL, Eapen GA, Ettinger DS, et al. Lung Cancer Screening, Version 3.2018, NCCN Clinical Practice Guidelines in Oncology. J Natl Compr Canc Netw. 2018; 16: 412–441.

16. Kubo T, Furuta T, Johan MP, Ochi M. Prognostic significance of18F-FDG PET at diagnosis in patients with soft tissue sarcoma and bone sarcoma; Systematic review and meta-analysis. Eur J Cancer. 2016; 58: 104–111.

17. Li YJ, Dai YL, Cheng YS, Zhang WB, Tu CQ. Positron emission tomography (18) F-fluorodeoxyglucose uptake and prognosis in patients with bone and soft tissue sarcoma: A meta-analysis. Eur J Surg Oncol. 2016; 42: 1103–1114.

18. Chen L, Wu X, Ma X, Guo L, Zhu C, et al. Prognostic value of 18F-FDG PET-CT-based functional parameters in patients with soft tissue sarcoma: A meta-analysis. Medicine (Baltimore). 2017; 96: e5913.