Case report

The early-stage of primary pulmonary synovial sarcoma: A case report

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ABSTRACT

Background: Primary pulmonary synovial sarcoma is a rare case of lung cancer. Surgical resection is standard therapy. Early-stage and complete resection offers a good prognosis.

Case presentation: A 50-year-old woman complained of intermittent left chest pain. Chest X-ray showed a mass around the heart. MSCT also showed enhanced solid lesion in the lower left lung anterobasal segment adhered to the pericardium. The patient underwent surgery with wedge resection with both immunohistochemical (IHC) EMA and CD99 positive. After surgery, she received 3-cycle chemotherapy that consisted of a combination of doxorubicin and ifosfamide.

Discussion: Surgical resection is the mainstay treatment for primary pulmonary synovial sarcoma. Chemotherapy can be considered for a patient with high-risk metastases. Early-stage and young age showed a good prognosis. Patient with stage IA still survives for 49 months after completing surgical resection and chemotherapy.

Conclusion: Resection of the tumor followed by chemotherapy may increase the survival tumor.

1. Introduction

Synovial sarcoma is a type of sarcoma more prevalent in men compared to women, accounting for 8% of all types of sarcoma. This sarcoma occurs due to the translocation t(X;18) (p11;q11) i.e. in chromosome 18 and X chromosomes the translocation takes place [1]. Primary pulmonary synovial sarcoma is very rare, accounting for <0.5% of all lung tumors, arising most commonly in the lung parenchyma followed by the pleura and mediastinum [2–4]. This type of tumor is very aggressive with poor clinical outcomes [2,5]. Primary pulmonary synovial sarcoma could occur in both genders equally [4,6]. In chest X-ray, synovial sarcoma often manifests as lung parenchyma mass with well-circumscribed rounded or lobulated borders [3,4,7]. Common computed tomography findings are solitary, large, well-circumscribed mass that enhances heterogeneously on contrast [2,7]. In this case, 51 years old female patient has a cough and chest pain. Chest CT scan showed enhanced solid mass in the left lower lobe. The patient was diagnosed with primary pulmonary synovial sarcoma and underwent surgery for left lower lobe resection and chemotherapy with a good response. This case reports primary pulmonary synovial sarcoma is based on SCARE guidelines 2020 [8].

2. Case presentation

A 50-year-old woman complained of intermittent left chest pain for one-month, dull pain, intermittent cough for two months, no phlegm, no coughing up blood, infrequent shortness of breath, no weight loss, and no fever. The patient had a history of hypertension and regularly takes anti-hypertension drugs, there was no history of cancer in the family. The investigation of the patient’s condition was good. Chest examination was normal. Laboratory examination was unremarkable and there was no axillary and cervical lymphadenopathy. There was no bump or pain due to pressure in the extremity and a clubbing finger was not found. The remaining systemic examination was normal. Laboratory examination was unremarkable. Supporting chest X-ray results showed a mass in the left lower lobe (Fig. 1).

Additional examination using multislice computed tomography (MSCT) of the thorax revealed enhanced solid lesions in the anterobasal segment of the left lobe with a size of 2.1 × 1.8 × 1.9 cm, adhered to the pericardium, enlarged subcentimeter lymph nodes in the right peribronchial, left peribronchial, sub carina, right and left lower paratracheal, and right upper paratracheal (Fig. 2). A bone survey also reveals no metastasis. FNAB has not carried into consideration of its near location to the heart. The patient underwent open biopsy with video-
assisted thoracoscopic surgery (VATS) procedure. Because of the difficulty to approach the tumor, it was decided to do thoracotomy. The tumor was found in the lower-left lobe with no adhesion to the surrounding tissue. Therefore, wedge resection with staples was conducted without lymphadenectomy. Extracted white, grey, dense, and springy mass with the size of $2.0 \times 1.5$ cm. Histopathological examination showed spindle cell tumor and synovial sarcoma diagnosis is based on the result of immunohistochemistry (IHC) epithelial membrane antigen (EMA) and CD 99 positive (Fig. 3) [4]. TNM staging is based on the American Joint Committee on Cancer (AJCC) eighth edition of bone and soft tissue sarcomas stage IA. T1 (organ-confined) based on the abdomen and thoracic visceral organs, N0 dan M0 [9].

Chemotherapy was then carried out for 3 cycles with doxorubicin at a dose of 25 mg/m$^2$ and ifosfamide at a dose of 3000 mg/m$^2$ along with mesna in three weekly protocols. Chest CT-scan evaluation was conducted three months after chemotherapy and showed complete response without local recurrence. The patient underwent regular check-ups every 3 months in the first-year post-surgery and adjuvant chemotherapy and every 6 months after that with a complete response without recurrence.

3. Discussion

The incident of primary pulmonary synovial sarcoma is very low. Patients mostly seek help due to cough, chest pain, difficulty in breathing, and hemoptysis. Although it resembles its soft tissue counterpart but is difficult to diagnose clinically and radiologically from other primary and metastatic lung tumors [7]. The diagnosis of primary pulmonary synovial sarcoma is based on clinical symptoms, radiological examination, pathology, and immunohistochemistry to exclude the possibility of another primary tumor and a metastatic sarcoma [10]. Immunohistochemical examination plays an important role in the diagnosis of primary pulmonary synovial sarcoma. Primary pulmonary synovial sarcoma is positive for cytokeratins 7 and 19, EMA, BCI-2, CD 99, and Vimentin, while negative for S-100, CD-34, desmin, actin, and tumor vascular markers. In addition to histopathological and immunohistochemical (IHC) examination, cytogenetic analysis can confirm the diagnosis of pulmonary synovial sarcoma [4,11].

The standard treatment of primary pulmonary synovial sarcoma is wide surgical There is still no standard guideline for adjuvant radiotherapy or chemotherapy, but radiotherapy is more recommended. Chemotherapy may be considered in patients at high risk of metastases.
or in advanced stages [11,12]. The patient underwent left lower lobe wedge resection followed by chemotherapy of doxorubicin and ifosfamide. Chemotherapy is an option because of the long waiting time. Doxorubicin and ifosfamide are the standard protocol for synovial sarcoma with expected response rates ranging from 25 up to 60% [11,13]. In addition, the combination of doxorubicin and ifosfamide has been shown to have a therapeutic effect in sarcomas [14].

In general, the prognosis for primary pulmonary synovial sarcoma is poor with an overall survival rate of 87% in 1 year and 59.4% in 5 years [15]. Primary intrathoracic synovial sarcoma is more aggressive, with an overall 5-year survival rate of 30% compared with 50% in soft tissue synovial sarcoma [16,17]. Age, tumor location, staging, and type of histopathology affect prognosis [11]. However, the main factor influencing the prognosis is complete resection [18,19]. Poor factors influencing tumor size (>5 cm), male sex, age (>20 years), extensive tumor necrosis, the number of mitotic figures (>10/10 high-powered fields), neurovascular invasion, and the SYT-SSX1 variant [4,16].

4. Conclusion

Primary pulmonary synovial sarcoma is a very rare tumor. Diagnosis and therapy are still a challenge. The early-stage has a good prognosis.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Provenance and peer review

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Ethical approval

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Fig. 3. A. Epithelial membrane antigen of positive; B. CD99 of positive; C. histopathology showed spindle cells.
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All authors contributed toward data analysis, drafting and revising the paper, gave final approval of the version to be published and agree to be accountable for all aspects of the work.

Declaration of competing interest

Diana Rahmaniar and Daniel Maranatha declare that they have no conflict of interest.

References

[1] N.Z. Ikeri, A.O. Akinjo, O.O. Ajayi, A.A. Fehintola Banjo, Histological types of soft tissue sarcomas at the Lagos university teaching hospital, Niger.Postgrad.Med.J. 24 (4) (2017) 205–209, https://doi.org/10.4103/npgm.npgm_146-17.

[2] B. Etienne-Mastroianni, L. Falchero, L. Chalabreysse, R. Loire, D. Ranchère, P. J. Seouquet, et al., Primary sarcomas of the lung: a clinicopathologic study of 12 cases, Lung Cancer (Amsterdam, Netherlands) 38 (3) (2002) 283–289, https://doi.org/10.1016/s0169-5002(02)00303-3.

[3] P.H. Hartel, J.C. Fanburg-Smith, A.A. Frazier, J.R. Galvin, J.H. Lichy, K. Shilo, et al., Primary pulmonary and mediastinal synovial sarcoma: a clinicopathologic study of 60 cases and comparison with five prior series, Mod. Pathol. 20 (7) (2007) 760–769, https://doi.org/10.1038/modpathol.3800790.

[4] M.K. Panigrahi, G. Pradhan, N. Sahoo, P. Mishra, S. Patra, P.R. Mohapatra, Primary pulmonary synovial sarcoma: a reappraisal, J. Cancer Res. Ther. 14 (3) (2018) 481–489, http://dx.doi.org/10.4103/jcrt.jcrt_1462-1804883.

[5] H.S. Sandeepa, A.H. Kate, P. Chaudhari, V. Chavan, K. Patole, N. Lokeshwar, et al., Primary pleural synovial sarcoma: a rare case of hemorrhagic pleural effusion in a young adult, J. Cancer Res. Ther. 9 (3) (2013) 517–519, https://doi.org/10.4103/0973-1482.119367.

[6] E.C. Seyhan, S.N. Sokucu, G. Gunluoglu, N.S. Veske, S. Altin, Primary pulmonary synovial sarcoma: a very rare presentation, Case Rep.Pulmonol. 2014 (2014), 537618, https://doi.org/10.1155/2014/537618.

[7] L. Pandey, D. Joseph, R. Panthick, M.K. Gupta, Primary synovial sarcoma of the lung: a rare presentation, diagnostic dilemma and review of literature, BMJ Case Rep. 13 (11) (2020), https://doi.org/10.1136/bcr-2020-237678.

[8] R.A. Agha, T. Franchi, C. Sohrabi, G. Mathew, A. Kerwan, The SCARE 2020 guideline: updating consensus Surgical CAse REport (SCARE) guidelines, Int.J.Surg.(London, England) 84 (2020) 225–230, https://doi.org/10.1016/j.ijsu.2020.10.034.

[9] K. Tanaka, T. Ozaki, New TNM classification (AJCC eighth edition) of bone and soft tissue sarcomas: JCOG Bone and Soft Tissue Tumor Study Group, Jpn. J. Clin. Oncol. 49 (2) (2019) 103–107, https://doi.org/10.1093/jjco/hyy157.

[10] R. Kumar, S. Menon, S.B. Desai, C.S. Pramesh, H. Menon, N.A. Jambhekar, Primary endobronchial synovial sarcoma confirmed by SYT-SSX1 fusion gene transcript by reverse transcriptase polymerase chain reaction, Indian J.Pathol.Microbiol. 52 (4) (2009) 520–523, https://doi.org/10.4103/0377-4929.56147.

[11] M. Fiore, A. Sambri, P. Spinnato, R. Zucchi, C. Giannini, E. Caldari, et al., The biology of synovial sarcoma: state-of-the-art and future perspectives, Curr. Treat. OptionsOncol. 22 (12) (2021) 109, https://doi.org/10.1007/s11864-021-09914-4.

[12] F.C. Eilber, S.M. Dry, Diagnosis and management of synovial sarcoma, J. Surg. Oncol. 97 (4) (2008) 314–320, https://doi.org/10.1002/jso.20974.

[13] M. Vlenterie, R.L. Jones, W.T. van der Graaf, Synovial sarcoma diagnosis and management in the era of targeted therapies, Curr. Opin. Oncol. 27 (4) (2015) 316–322, https://doi.org/10.1097/cco.0000000000000198.

[14] S. Verma, J. Younus, D. Stry-Norman, A.E. Haynes, M. Blackstein, Meta-analysis of ifosfamide-based combination chemotherapy in advanced soft tissue sarcoma, Cancer Treat. Rev. 34 (4) (2008) 339–347, https://doi.org/10.1016/j.ctrv.2008.01.005.

[15] M.N. Aytekin, R. Oztürk, K. Amer, A. Yapur, Epidemiology, incidence, and survival of synovial sarcoma subtypes: SEER database analysis, J.Orthop.Surg.(Hong Kong) 28 (2) (2020), 2309499020936009, https://doi.org/10.1177/2309499020936009.

[16] S. Dennison, E. Weppler, G. Giacoppe, Primary pulmonary synovial sarcoma: a case report and review of current diagnostic and therapeutic standards, Oncolgoc 9 (3) (2004) 339–342, https://doi.org/10.1634/thoracolgoc.9-3-339.

[17] R.F. Falkenstern-Ge, M. Kimmich, A. Grabner, H. Horn, G. Friedel, G. Ott, et al., Primary pulmonary synovial sarcoma: a rare primary pulmonary tumor, Lung 192 (1) (2014) 211–214, https://doi.org/10.1007/s00408-013-9521-1.

[18] M. Trassard, V. Le Dousal, K. Hactene, P. Terrier, D. Ranchère, L. Guillou, et al., Prognostic factors in localized primary synovial sarcoma: a multicenter study of 128 adult patients, J.Clinc.Oncol. 19 (2) (2001) 523–534, https://doi.org/10.1200/jco.2001.19.2.525.

[19] H. Zhang, W. Huang, Q. Feng, W. Sun, W. Yan, C. Wang, et al., Clinical significance and risk factors of local recurrence in synovial sarcoma: a retrospective analysis of 171 cases, Front.Surg. 8 (2021), 736146, https://doi.org/10.3389/fsurg.2021.736146.