



Case report

A female with solitary fibrous tumor pleura mimicking pulmonary sclerosing pneumocytoma in low resource setting: A case report

Sakina^a, Isnin Anang Marhana^{a,*}, Dhihintia Jiwangga^b

^a Department of Pulmonology and Respiratory Medicine, Faculty of Medicine, Universitas Airlangga – Dr. Soetomo General Academic Hospital, Surabaya, Indonesia

^b Department of Thoracic and Cardiovascular, Surgery, Faculty of Medicine, Universitas Airlangga – Dr. Soetomo General Academic Hospital, Surabaya, Indonesia

ARTICLE INFO

Keywords:

Pulmonary sclerosing pneumocytoma
Solitary fibrous tumors of pleura
Thoracotomy with intercostal incision

ABSTRACT

Background: Solitary fibrous tumor of pleura (SFTP) is a rare condition. Clinical symptoms and non-specific radiological features in both tumors make preoperative diagnosis difficult to establish.

Case presentation: A Javanese 47-year-old female complained of chest pain and shortness of breath which an X-ray and CT-Scan of the thorax showed giant cell metastases in the lung. Signs and symptoms indicate pulmonary sclerosing pneumocytoma (PSP) and the patient underwent thoracic surgery which was first performed with angiographic embolization. Furthermore, an anatomical pathology analysis was performed with suspected SFTP, supported by the IHC test, which found CD34 (+), EMA (–), and S100 (–). Thoracic surgery followed by radiotherapy and chemotherapy is recommended in SFTP patients.

Discussion: The SFTP and PSP have non-specific clinical symptoms and radiological features. Anatomic pathology and IHC test are definitive diagnostic tools from SFTP and PSP. Establishing a preoperative diagnosis of SFTP and PSP is quite difficult. Surgical resection is the treatment of choice for both.

Conclusion: The comparison of SFTP and PSP can be seen results of the anatomical pathology analysis and IHC test.

1. Introduction

Primary pleural tumors are benign or malignant lesions arising from either the parietal or visceral pleura. The most common of non-mesothelioma primary pleural tumors are solitary fibrous tumors of pleura (SFTP) [1]. In North America, it is estimated that tumor incidence is about 2.8 cases per 100,000 hospitalizations. Malignant mesothelioma is more than 90% of primary pleural tumors. Of the remaining 10%, about 5% are classified as SFTP and the other 5% are various less common primary pleural tumors [2]. Moreover, pleural tumors cause 0.3% to 3.5% thoracic tumors globally [1,3]. The preoperative differential diagnosis that occurs in patients with SFTP is any mass lesion in the chest, ranging from pulmonary carcinoma to various intrapleural sarcomas [4]. In low resource setting, misdiagnosis is possible between SFTP and pulmonary sclerosing pneumocytoma (PSP) [5] because of the similarity of signs and symptoms and limited investigations [6,7]. Based on the description above, we are interested in reporting a case of solitary fibrous tumors of pleura using the surgical case report (SCARE) 2020 guideline [8].

2. Case presentation

A Javanese 47-years-old female complained of chest pain and shortness of breath since 3 months ago. Patients experienced significant weight loss without any decrease in appetite. The patient was suspected of having a lung tumor based on physical and radiological examination (Fig. 1A) when the patient went to the emergency department for asthma exacerbations. Patients were diagnosed with asthma since childhood. Thoracic CT scan with contrast appeared to be a solid lesion (39 HU) which by contrast was increasingly enhanced in the next phase (133 HU). The tumor size of $\pm 10.7 \times 14.1 \times 9.72$ cm in the upper left hemorrhage was attached to the pleura (Fig. 1B). The examination of fiber optic bronchoscopy (FOB) showed a blunt primary carina, narrowing the distal lumen of the main bronchial due to pressure from the posterior wall, narrowing the left main bronchial lumen caused by the pressure of extra lumen mass. The FOB obtained material for anatomic pathology examination and the results showed no malignant cells (Fig. 2).

Embolization was performed, showing the hypervascular mass in the

* Corresponding author at: Isnin Anang Marhana, Department of Pulmonology and Respiratory Medicine, Faculty of Medicine, Universitas Airlangga – Dr. Soetomo General Academic Hospital, Jl. Mayjend Prof. Dr. Moestopo No. 6-8, Airlangga, Gubeng, Surabaya, East Java 60286, Indonesia.

E-mail address: isnin.anang@fk.unair.ac.id (I.A. Marhana).

<https://doi.org/10.1016/j.ijscr.2022.106928>

Received 30 December 2021; Received in revised form 8 February 2022; Accepted 5 March 2022

Available online 9 March 2022

2210-2612/© 2022 The Authors. Published by Elsevier Ltd on behalf of IJS Publishing Group Ltd. This is an open access article under the CC BY-NC-ND license

(<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

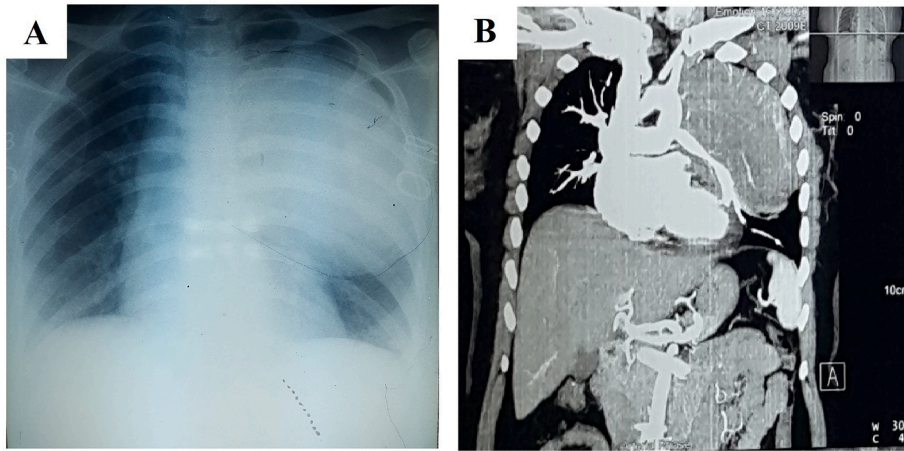


Fig. 1. A) X-ray and B) preoperative CT-Scan of the lung.

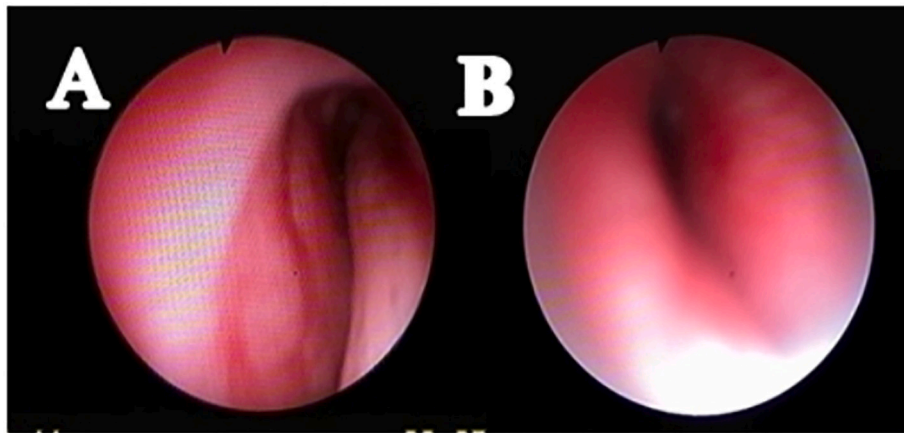


Fig. 2. A) Right main bronchus and B) left main bronchus.

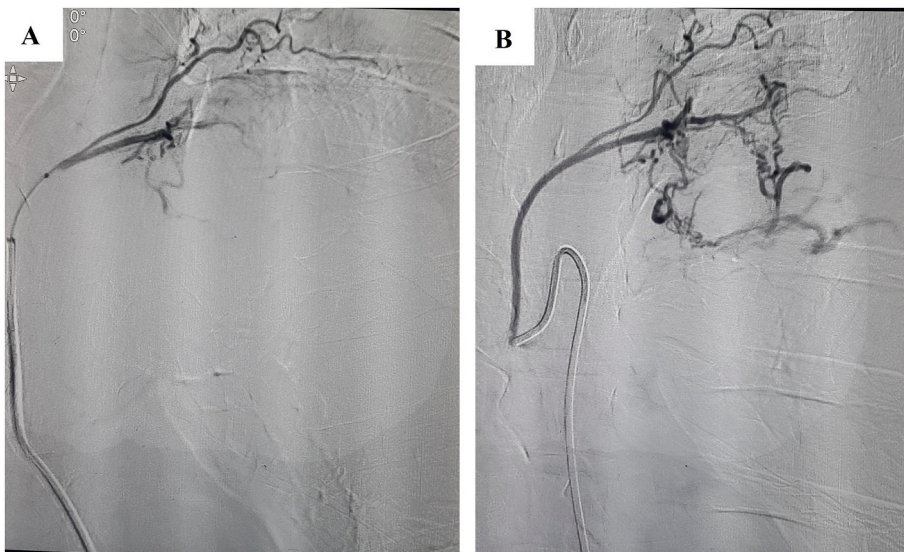


Fig. 3. Arteriography results recanalized the left bronchial artery.

left hemithorax which fed on the left intercostal artery level Th 5–6, the recanalized left bronchial artery, and fine branches of other intercostal artery levels. Post vascularization embolization from the feeding arteries of the superior branches of the left intercostal artery was completely

closed. Feeding of the recanalized left bronchial artery and fine branches of other intercostal arteries could not be embolized (Fig. 3).

The surgical technique uses an anterolateral thoracotomy approach and enters through the 5th intercostal space. The tumor was seen as a

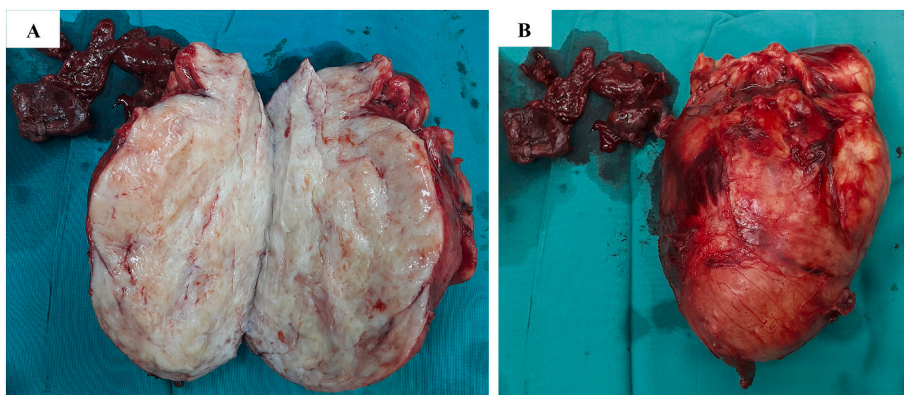


Fig. 4. Size tumor 12 × 8 × 5 cm.

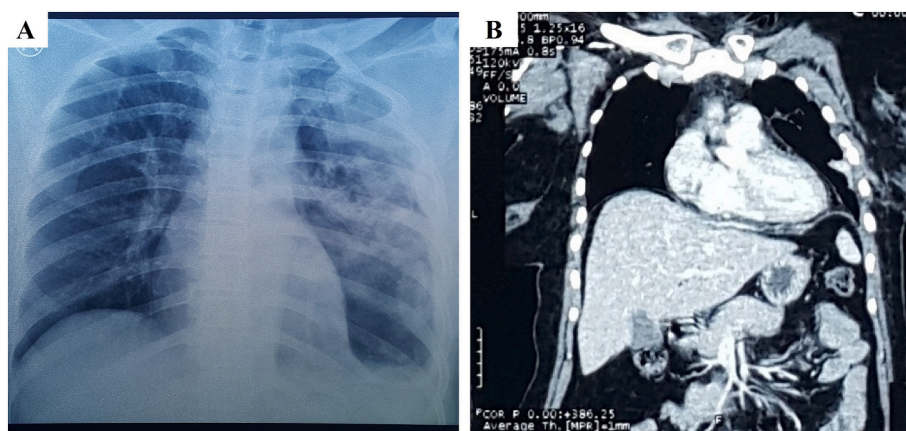


Fig. 5. Postoperative radiological examination.

round, well-defined tumor, mounted on the posterior thoracic wall with a size of 12 × 8 × 5 cm (Fig. 4) which anatomical pathology analysis showed tissue sections arranged solidly in irregular patterns, consisting of proliferating oval-spindle-nucleated cells and fine chromatin (SFTP). The results of the immunohistochemical (IHC) test are epithelial membrane antigen (EMA) negative in the membrane and cytoplasm of tumor cells, S100 negative in the cytoplasm of tumor cells, and the cluster of differentiation 34 (CD34) positive in tumor cell membranes. It was concluded that the tumor was a SFTP. A radiological examination was performed to evaluate the patient, showing no mass found in the right/left lung/mediastinum (Fig. 5). After surgery, the patient did not feel any complaints. The patient underwent radiotherapy and chemotherapy. One-year postoperative evaluation, the patient had no respiratory complaints and no clinical or radiological signs of recurrence were found.

3. Discussion

A solitary fibrous pleural tumor is a primary tumor arising from the pleura that can be a benign tumor and has various levels to potentially become malignant [1]. There are two types of primary pleural tumors, namely diffuse and localized. Localized tumors are rare, known as SFTP, which arise from the sub-mesothelial mesenchymal layer. An SFTP occurs in a wide age range (5 to 87 years), often found to occur at the age of the sixth and seventh decades, with quite the same frequency in both sexes [4]. The diagnosis of SFTP and PSP can be distinguished, one of them by using dynamic CT-Scan which dynamic CT-Scan can provide a more accurate picture of the lesion in PSP cases [9]. Dynamic CT-Scan was not performed in our case due to resource constraints. Based on

X-ray and CT-scan, giant malignancy is not specific. Support argument diagnosis of SFTP base FOB with malignancy results. The IHC test in SFTP included a primary positive CD34 result, followed by CD99, cytokeratin (S100), and epithelial membrane antigen (EMA) [10]. CD34 is the most frequently used biomarker for SFTP, which in PSP patients the IHC examination was seen for TTF-1, EMA, and pan-cytokeratin immunostaining [11–13].

In cases of SFTP, surgery is the preferred action which in cases of malignancy can be surgical procedures performed safely without a preoperative diagnosis [11]. The surgical procedure chosen was thoracotomy with an intercostal incision which is a surgical technique that is effectively removing the giant tumor in the lung [14]. Before surgery, angiographic embolization is now widely accepted to reduce the occurrence of intraoperative bleeding. Embolization is usually performed 24–72 h before resection to help hemostasis during surgery [15]. Post-surgery, patients are at risk for pain and atelectasis so they need to get special attention for pain management and physiotherapy [16]. Meanwhile, some literature recommends that patients undergo radiotherapy or chemotherapy post-surgery [17,18].

4. Conclusion

A Javanese 47-years-old female complained of chest pain and shortness of breath which an X-ray and CT-Scan of the thorax showed giant cell metastases in the lung. Signs and symptoms indicate PSP and the patient underwent thoracic surgery which was first performed with angiographic embolization. Furthermore, an anatomical pathology analysis was performed with suspected SFTP, supported by the IHC test, which found CD34 (+), EMA (–), and S100 (–). Thoracic surgery

followed by radiotherapy and chemotherapy is recommended in SFTP patients. The comparison of SFTP and PSP can be seen results of the anatomical pathology analysis and IHC test.

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.

Guarantor

Isnin Anang Marhana is the person in charge of the publication of our manuscript.

Provenance and peer review

Not commissioned, externally peer-reviewed.

Sources of funding

None.

Ethical approval

Not applicable.

Research registration

Not applicable.

Credit authorship contribution statement

Sakina: Data curation, supervision, visualization, investigation, and drafting; **Isnin Anang Marhana:** Conceptualization, methodology, drafting, editing, revising, and reviewing; **Dhahintia Jiwangga:** Methodology, editing, and revising.

Declaration of competing interest

Sakina, Isnin Anang Marhana, and Dhahintia Jiwangga Suta Winarno declare that they have no conflict of interest.

Acknowledgment

We would like to thank our editor "Fis Citra Ariyanto".

References

- [1] C.T. Erb, K.M. Johnson, A.W. Kim, Rare pleural tumors, *Clin. Chest Med.* 34 (1) (2013) 113–136, <https://doi.org/10.1016/j.ccm.2012.12.001>.
- [2] M.J. Chung, K.S. Lee, J. Han, Y.M. Sung, S. Chong, O.J. Kwon, Pulmonary sclerosing hemangioma presenting as solitary pulmonary nodule: dynamic CT findings and histopathologic comparisons, *Am. J. Roentgenol.* 187 (2) (2006) 430–437, <https://doi.org/10.2214/ajr.05.0460>.
- [3] H. Yagyu, Y. Hara, K. Murohashi, Y. Ishikawa, T. Isaka, T. Woo, et al., Giant solitary fibrous tumor of pleura presenting both benign and malignant features, *Am. J. Case Rep.* 20 (2019) 1755–1759, <https://doi.org/10.12659/ajcr.919639>.
- [4] L.A. Robinson, Solitary fibrous tumor of the pleura, *Cancer Control* 13 (4) (2006) 264–269, <https://doi.org/10.1177/107327480601300403>.
- [5] B. Yalcin, T.T. Bekci, S. Kozacioglu, O. Bolukbas, Pulmonary sclerosing pneumocytoma, a rare tumor of the lung, *Respir. Med. Case Rep.* 26 (2019) 285–287, <https://doi.org/10.1016/j.rmcr.2019.02.002>.
- [6] R. Sullivan, O.I. Alatisse, B.O. Anderson, R. Audisio, P. Autier, A. Aggarwal, et al., Global cancer surgery: delivering safe, affordable, and timely cancer surgery, *Lancet Oncol.* 16 (11) (2015) 1193–1224, [https://doi.org/10.1016/s1470-2045\(15\)00223-5](https://doi.org/10.1016/s1470-2045(15)00223-5).
- [7] P.A. Cowper, L. Feng, A.S. Kosinski, B.C. Tong, R.H. Habib, J.B. Putnam Jr., et al., Initial and longitudinal cost of surgical resection for lung cancer, *Ann. Thorac. Surg.* 111 (6) (2021) 1827–1833, <https://doi.org/10.1016/j.athoracsur.2020.07.048>.
- [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. (Lond. Engl.)* 84 (2020) 226–230, <https://doi.org/10.1016/j.ijsu.2020.10.034>.
- [9] H.Y. Le, D.P. Pham, K.T. Nguyen, V.A. Hoang, T.S. Trinh, Q. Do, Pulmonary sclerosing pneumocytoma in an 18-year-old male patient: a case report and literature review, *Medicine* 99 (26) (2020), e20869, <https://doi.org/10.1097/md.00000000000020869>.
- [10] R.J. Vogels, M. Venterie, Y.M. Versleijen-Jonkers, E. Ruijter, E.M. Bekers, M. A. Verdijk, et al., Solitary fibrous tumor - clinicopathologic, immunohistochemical and molecular analysis of 28 cases, *Diagn. Pathol.* 9 (2014) 224, <https://doi.org/10.1186/s13000-014-0224-6>.
- [11] L. Zhou, C. Sun, Y. Huang, Q. Li, H. Tang, Y. Wang, Pulmonary sclerosing hemangioma with a rare symptom: a case report and review of the literature, *Mol. Clin. Oncol.* 6 (2) (2017) 221–224, <https://doi.org/10.3892/mco.2016.1114>.
- [12] B. Chen, J. Gao, H. Chen, Y. Cao, X. He, W. Zhang, et al., Pulmonary sclerosing hemangioma: a unique epithelial neoplasm of the lung (report of 26 cases), *World J. Surg. Oncol.* 11 (2013) 85, <https://doi.org/10.1186/1477-7819-11-85>.
- [13] N.J. Olson, K. Linos, Dedifferentiated solitary fibrous tumor: a concise review, *Arch. Pathol. Lab. Med.* 142 (6) (2018) 761–766, <https://doi.org/10.5858/arpa.2016-0570-RS>.
- [14] M. Yanagiya, J. Matsumoto, T. Miura, H. Horiuchi, Extended thoracotomy with subcostal incision for giant solitary fibrous tumor of the diaphragm, *AME Case Rep.* 1 (2017) 8, <https://doi.org/10.21037/acr.2017.11.02>.
- [15] A. Panda, A.S. Bhalla, A. Goyal, Bronchial artery embolization in hemoptysis: a systematic review, *Diagn. Intervent. Radiol.* 23 (4) (2017) 307–317, <https://doi.org/10.5152/dir.2017.16454>.
- [16] R. Bar-Haim, A. Gavrilo, A. Samokhvalov, E. Altman, Solitary fibrous tumour: a rare tumour of the pleural cavity, *BMJ Case Rep.* 2017 (2017), <https://doi.org/10.1136/bcr-2016-217880>.
- [17] R.M. Mercer, C. Wigston, R. Banka, G. Cardillo, R. Benamore, A.G. Nicholson, et al., Management of solitary fibrous tumours of the pleura: a systematic review and meta-analysis, *ERJ Open Res.* 6 (3) (2020), <https://doi.org/10.1183/23120541.00055-2020>.
- [18] W.L. Liu, W. Wu, Q.C. Hong, K. Lv, Recurrence rates of surgically resected solitary fibrous tumours of the pleura: a systematic review and meta-analysis, *Interact. Cardiovasc. Thorac. Surg.* 32 (6) (2021) 882–888, <https://doi.org/10.1093/ivctv/ivab012>.