American Journal of Case Reports

e-ISSN 1941-5923
© Am J Case Rep, 2023; 24: e938696
DOI: 10.12659/AJCR.938696

Received: 2022.10.19
Accepted: 2022.12.07
Available online: 2022.12.13
Published: 2023.01.04

A Case of Primary Epithelioid Sarcoma of the Pleura

Authors' Contribution:
Study Design A
Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
Literature Search F
Funds Collection G

ABCDEF **Takashi Yamashita**

Department of General Thoracic and Breast Surgery, Iwata City Hospital, Iwata, Shizuoka, Japan

Corresponding Author: Takashi Yamashita, e-mail: takashiyamashita.md@gmail.com, takashiyamashita@hospital.iwata.shizuoka.jp
Financial support: None declared
Conflict of interest: None declared

Patient: Female, 64-year-old
Final Diagnosis: Epithelioid sarcoma
Symptoms: Dyspena
Medication: —
Clinical Procedure: —
Specialty: Oncology

Objective: Rare disease

Background: Epithelioid sarcoma is a rare tumor and that is extremely rare as a primary pleural neoplasm. On imaging, it may appear similar to malignant pleural mesothelioma; thus, it can be difficult to diagnose.

Case Report: A 64-year-old Asian woman, who had a treatment history of cervix adenocarcinoma, was admitted with dyspnea and right massive pleural effusion. Chest drainage was performed, and malignant cells were found in the pleural effusion. The malignant cells were thought to be metastasized from previous cervical cancer. We continued pleural drainage; however, the volume of the pleural effusion did not decrease. On the 5th hospital day, the chest tube became occluded. Computed tomography showed structures similar to empyema. Pleural irrigation and fibrinolytic therapy did not improve her condition. Empyema curettage was performed on the 14th hospital day. The resected pleura was submitted for pathological examination and showed tumor lesion but not metastatic adenocarcinoma of the cervix. The intrathoracic tumor grew extremely rapidly, and the patient died of respiratory failure on postoperative day 8 (22nd hospital day) before a diagnosis could be made. The final pathological diagnosis obtained on the 34th hospital day was epithelioid sarcoma.

Conclusions: For patients who appear to have empyema complicated by neoplastic lesions, a histopathological examination should also be performed to ensure accurate diagnosis. In addition, if a tumorous lesion is detected and it is neither metastatic nor malignant pleural mesothelioma, pleural epithelioid sarcoma should be added to the differential diagnosis in the presence of a rapidly growing and histologically difficult-to-diagnose pleural tumor.

Keywords: Empyema, Pleural • Sarcoma • SMARCB1 protein, human

Full-text PDF: https://www.amjcaserep.com/abstract/index/idArt/938696

Indexed in: [PMC] [PubMed] [Emerging Sources Citation Index (ESCI)] [Web of Science by Clarivate]

Background

Epithelioid sarcoma (ES), a malignant tumor with mixed differentiation into both mesenchymal and epithelial cells, was first reported by Enzinger in 1970 [1]. There is wide variation in the 5- year survival rates, ranging from 25% to 78% [2]. It is divided into 2 subtypes: the classic subtype, which occurs distally in the extremities; and the proximal-type subtype, which occurs in the proximal extremities and trunk [3]. The proximal-type subtype has been reported to occur in the perineum, pubis, genitalia, and trunk [4], but only a few cases of ES of pleural origin have been reported. ES in a rare site in which it is difficult to make a definitive diagnosis, but it was reported that ES was characterized by INI-1 deletion in approximately 90% of cases [5].

We experienced a case of proximal-type pleural ES (PES) that grew rapidly. It is difficult to distinguish from malignant pleural mesothelioma (MPM) based on imaging alone, and MPM is relatively difficult to diagnose histologically. Therefore, pathological diagnosis of PES was very difficult because of its low incidence, and the patient died while waiting for diagnosis. However, if a rapidly growing pleural tumor is suspected to be PES, it can be diagnosed by confirming the presence of INI-1 deletion. Herein, we report a rare case of PES and its clinical course.

Case Report

A 64-year-old Asian woman was referred to the respiratory division of our hospital for dyspnea on exertion. Her past medical history was adenocarcinoma of the cervix at the age of 63 years, and concurrent chemoradiotherapy performed 4 months prior to consultation showed partial response. She drank alcohol occasionally and did not have a history of cigarette smoking or illicit substance use.

She had had a cough for a week. Her dyspnea had worsened since the morning and she was referred to the emergency department. Her right respiratory sound was weakened and a chest X-ray showed a massive right pleural effusion. A chest tube was inserted into her right pleural cavity (**Figure 1**). Chest fluid examination showed low pH (7.00), low glucose (27 mg/dL), high lactate dehydrogenase (1950 U/L), high white blood cells (over the inspection limit), normal carcinoembryonic antigen (2.8 ng/mL), normal hyaluronic acid (234 000 ng/mL), and normal adenosine deaminase (22.8 IU/L). Additionally, cytology revealed malignant cells and culture showed no bacterial infection. Bacterial infection was not proven, but empyema was diagnosed comprehensively based on other examinations. Antibiotics were administered and we repeatedly drained and clamped the pleural effusion at a rate of 1000 mL/day to avoid re-expansion pulmonary edema, but it accumulated daily.

On the 5th hospital day, chest fluid stopped increasing and the chest tube seemed to be occluded. Computed tomography (CT) revealed many encapsulated fluid cavities similar to empyema (**Figure 2**). At the same time, a rapid thickening of the pleura was also observed, which was considered to be an inflammatory fibrin clot. An additional chest tube was inserted and pleural irrigation and intrapleural fibrinolytic therapy with urokinase were performed for 4 consecutive days; however, her symptoms did not improve. Our division was consulted, and we planned curettage of the cavity and pleurodesis.

We operated on the 14th hospital day. The encapsulated cavity was curetted, and thickened pleura was resected via

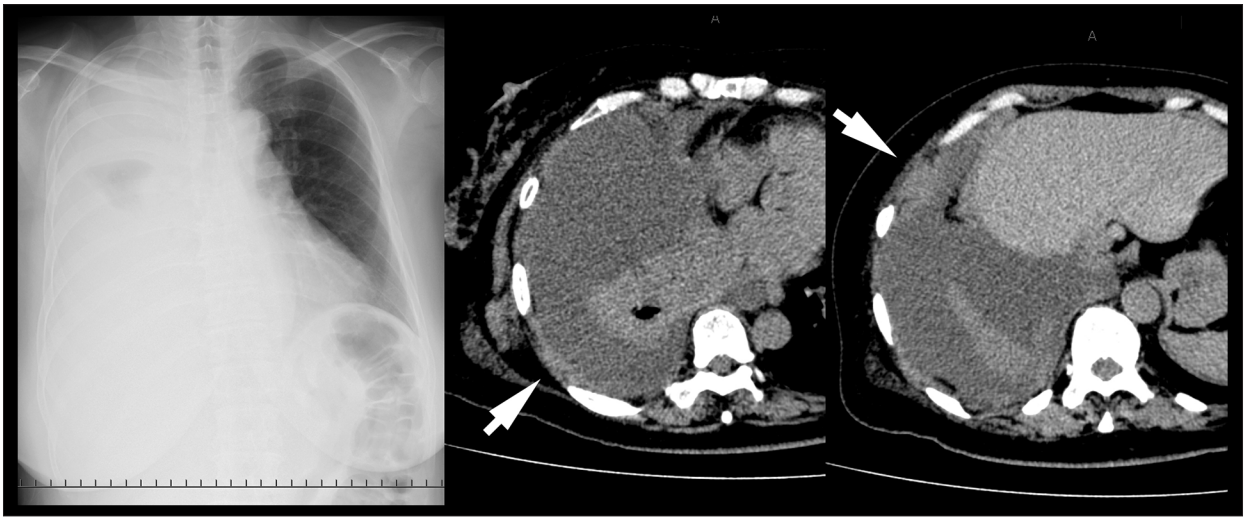

Figure 1. Chest X-ray and CT on the 1st hospital day. Chest X-ray showing right massive pleural effusion. CT showing right massive pleural effusion and collapse of the right lung. Partial pleural thickening can be noted retrospectively (white arrow).

Indexed in: [PMC] [PubMed] [Emerging Sources Citation Index (ESCI)] [Web of Science by Clarivate]

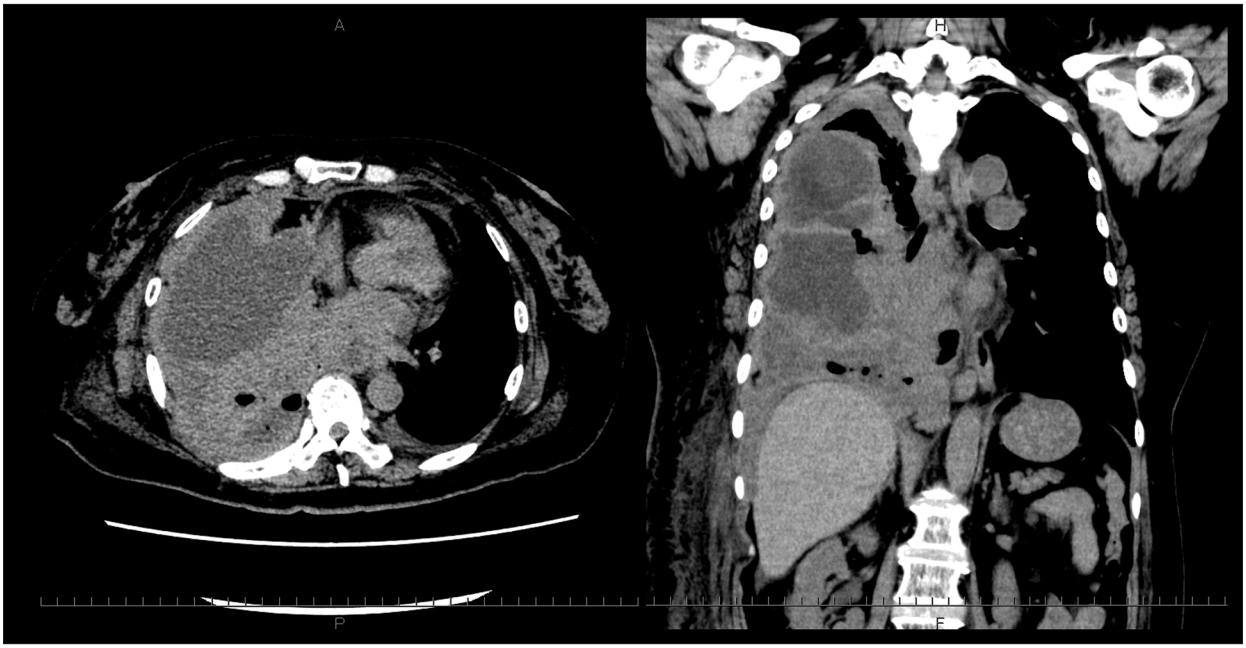

Figure 2. CT on 8th hospital day. CT showing many encapsulated fluid cavities in right thoracic cavity. It showed inflammation in the thoracic cavity, suggesting empyema.

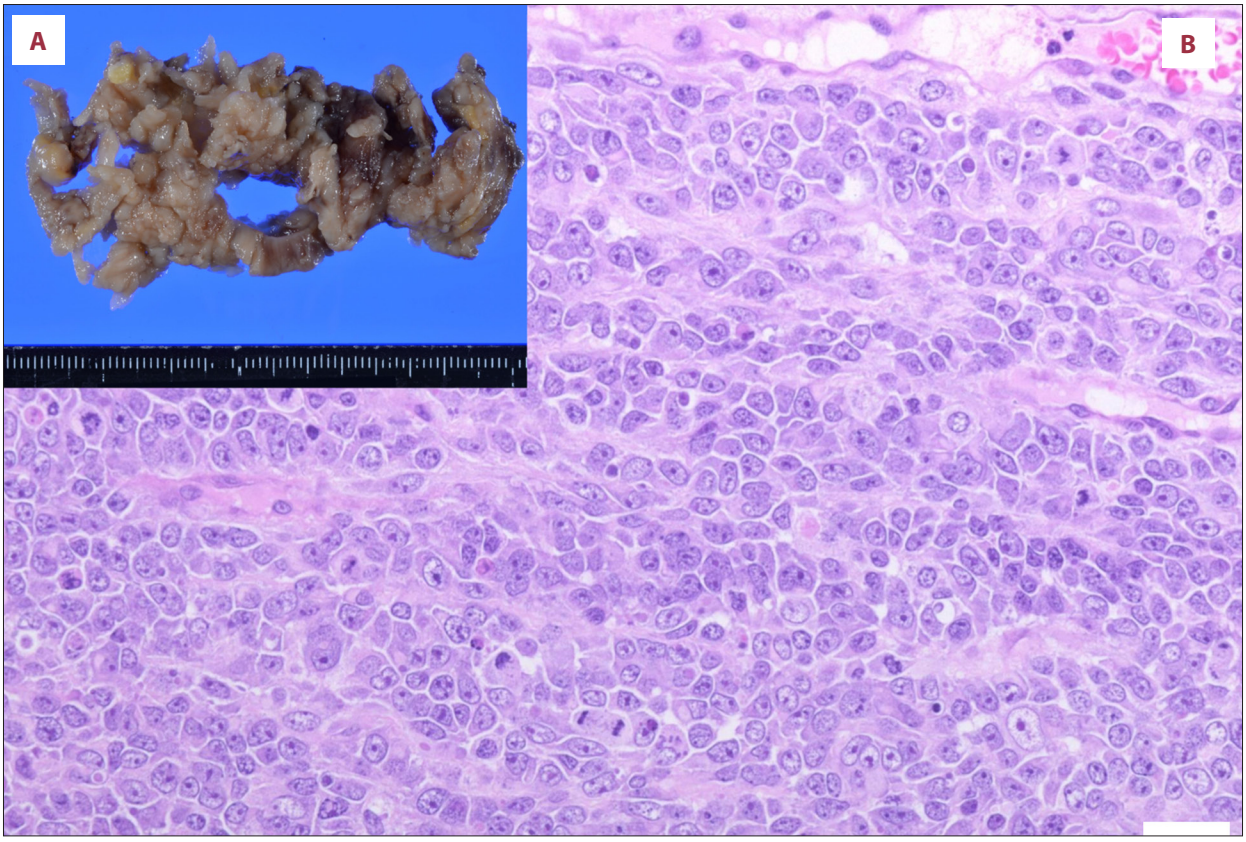

Figure 3. Resected tumor and pathological findings. (**A**) A portion of the resected tumor was submitted for pathological diagnosis. The specimen was thickened, elastic, and hard, like a tumorous lesion rather than inflammatory fibrin tissue. (**B**) Tumor cells showing an undifferentiated morphology with an oval nucleus with increased chromatin and a narrow cytoplasm are observed.

Indexed in: [PMC] [PubMed] [Emerging Sources Citation Index (ESCI)] [Web of Science by Clarivate]

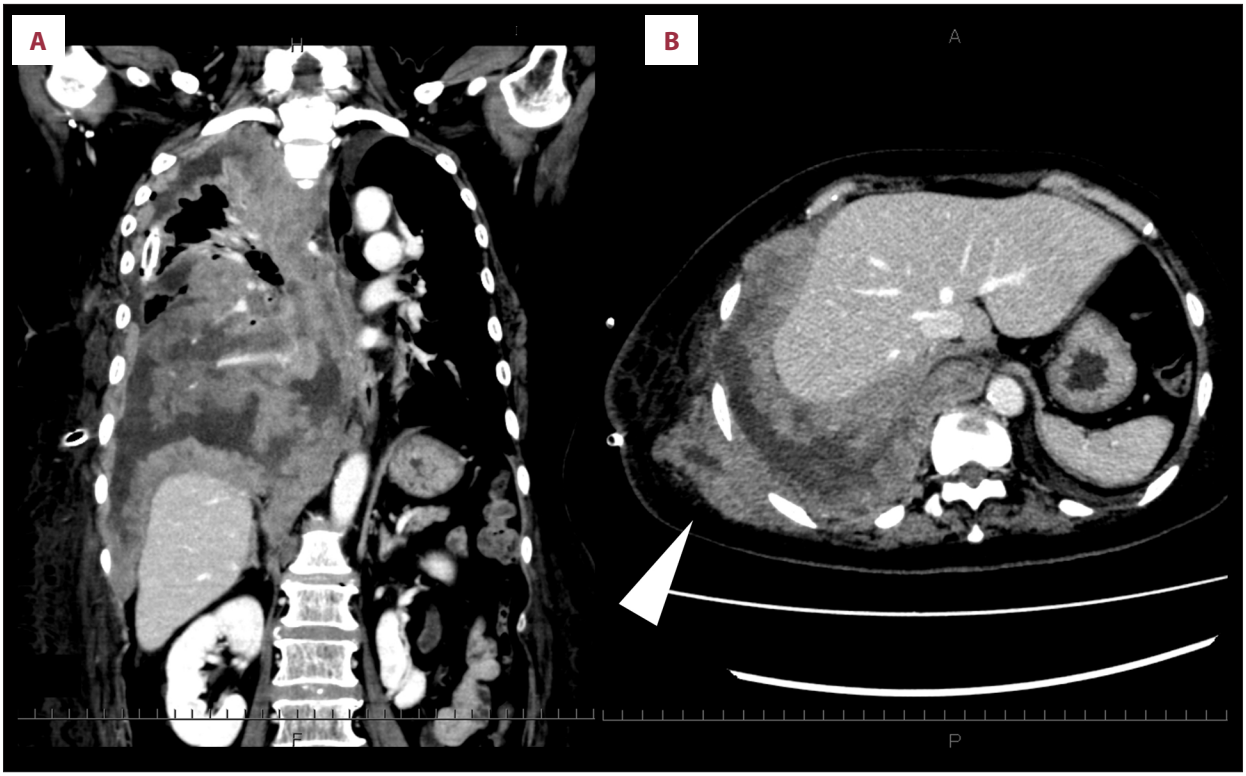

Figure 4. Axial and coronal enhanced CT on 19th hospital day (POD 5). (**A**) Although the thoracic cavity was curetted, new pleural thickening was observed. A thick tumorous growth was seen, especially on the diaphragm, which was hardly curetted in surgery. The left thoracic cavity was compressed due to rapid growth of the tumor. (**B**) Growth of disseminated tumor tissue was observed in the subcutaneous tissue at the site of initial chest drainage (white arrowhead).

thoracotomy (**Figure 3**). Because the resected pleura was thickened, elastic, and hard, like a tumorous lesion rather than inflammatory fibrin tissue, it was submitted for pathological examination. We could not perform pleurodesis because of circulatory instability induced by bleeding. After surgery, she was received a blood transfusion, and her general condition was stabilized until postoperative day (POD) 1. Intraoperative effusion culture showed no bacterial infection. The pathologists reported that most of the resected pleura were tumor lesions but not metastatic adenocarcinomas of the cervix. Based on the report, CT from the neck to the pelvis was performed again, but we observed no clear primary lesion. In addition, no obvious abnormalities were found in a physical examination that included the crown, extremities, and genitals. As we had difficulty in diagnosing her pathologically, we examined immunohistochemical staining of the tumor in stepwise fashion. Accordingly, immunohistological examination was positive for cytokeratin AE1/AE3, vimentin, EMA (partial), CD34 (partial), and β-catenin (membranous and focal cytoplasmic). Staining for INI1, calretinin, TTF-1, chromogranin A, synaptophysin, STAT6, WT1, Melan A, HMB-45, S-100, and NUT were negative. As a result of repeated discussions with 2 pathologists, INI1 negativity was an important decisive factor; finally, we diagnosed ES of the pleura on POD 20 (34th hospital day). Her right lung was not expanded, despite curettage and manual inflation during surgery. Her respiratory disorder improved temporarily after the operation as the pleural effusion was drained, but her general condition weakened day to day because of extremely rapid tumor progression (**Figure 4**). She did not wish to receive intubated ventilation after hearing that the respiratory failure caused by the tumor did not improve until treatment was started. On POD7, she developed CO_2 narcosis and became unconscious. She regained consciousness with manual mask ventilation, but when she was put on NPPV, she refused to put it on because of discomfort. She stated that if CO_2 narcosis redeveloped, she wanted only symptom relief and desired no treatment. We planned to perform anti-tumor treatment immediately after diagnosis; however, CO_2 narcosis redeveloped and she died on POD 8 (the 22nd hospital day) because of a respiratory disorder. The family did not provide consent for a pathological autopsy.

Discussion

Epithelioid sarcoma is rare, accounting for less than 1% of soft tissue sarcomas [6]. It is immunohistochemically positive for cytokeratin and vimentin, both mesenchymal and epithelial

Indexed in: [PMC] [PubMed] [Emerging Sources Citation Index (ESCI)] [Web of Science by Clarivate]

differentiation are observed, and INI-1 deletion is characteristic. Of the proximal-type, only 2 cases of pleural origin have been reported [7,8]. Previous lung cancer studies reported that the average solid-part tumor volume doubling time is 394~458 days [9,10]. In the present case, the volume of the tumor that had disseminated outside the thorax due to drainage was measured using the SYNAPSE VINCENT 3D volume analyzer (Fuji Medical Systems, Tokyo, Japan). The volume doubling time was calculated by the Schwartz equation [11] as approximately 7 days. This indicates an extremely rapid progression rate. Such a fast progression has not been reported in proximal-type ES and other tumors.

In the present case, it was believed that the marked imaging changes were due to empyema, even though the results of chest fluid aspiration indicated no evidence of bacterial infection. The rapid growth of a tumor causing carcinomatous pleurisy is uncommon; however, in the present case, a diagnosis of carcinomatous pleurisy was made. Therefore, there was an opportunity to suspect a cancerous lesion and a histological examination was performed at the time of surgery. During surgical procedures for typical empyema, cultures are often collected, but histological examination is not common. However, if surgical treatment is to be performed, a histopathological examination should also be performed to ensure accurate diagnosis, especially in cases that appear complicated by neoplastic lesions. In this patient, positive cytology became an indication for tissue diagnosis.

Most tumors growing in the pleura are MPM or metastatic tumors, or rarely a lymphomatous tumor [12]. Differential diagnosis is extremely difficult when none of the above is diagnosed. Additionally, MPM is relatively difficult to diagnose histologically, even immunohistochemically; therefore, MPM is occasionally diagnosed clinically without characteristic findings. On the other hand, this case suggested PES as a possible disease of rapidly growing pleural tumor. The progression of most MPM is not as rapid as PES. Since it is difficult to distinguish PES from MPM based on imaging alone, it is recommended that PES should also be suspected, especially in the presence of a histologically difficult-to-diagnose, rapidly growing pleural tumor. Diagnosis of PES may be possible by examination of INI-1 deletion if it is included in the differential diagnosis.

Conclusions

In cases that appear to be empyema, there is a slight possibility of a rapid growing tumor. Therefore, a histopathological examination should also be performed to ensure accurate diagnosis, especially in patients who seem to be complicated by neoplastic lesions. In addition, if a tumorous lesion is detected and it is neither metastatic nor MPM, PES should be added to the differential diagnosis in the presence of a rapidly growing and histologically difficult-to-diagnose pleural tumor.

Acknowledgements

I thank Dr. Shioto Suzuki and Dr. Ippei Ohnishi, who are staff of the Division of Diagnostic Pathology, for examination and making the diagnosis.

Declaration of Figures' Authenticity

All figures submitted have been created by the authors who confirm that the images are original with no duplication and have not been previously published in whole or in part.

References:

1. Enzinger FM. Epithelioid sarcoma. A sarcoma simulating a granuloma or a carcinoma. Cancer. 1970;26:1029-41
2. de Visscher SA, van Ginkel RJ, Wobbes T, et al. Epithelioid sarcoma: Still an only surgically curable disease. Cancer. 2006;107:606-12
3. Oda Y, Dal Cin P, Le Loarer F, Nielsen TO. Epithelioid sarcoma. In: WHO Classification of Tumours Editorial Board. WHO Classification of Tumours, 5th Ed., Vol. 3 Soft Tissue & Bone Tumours. Lyon: IARC; 2020;294-96
4. Guillou L, Wadden C, Coindre JM, et al. "Proximal-type" epithelioid sarcoma, a distinctive aggressive neoplasm showing rhabdoid features. Clinicopathologic, immunohistochemical, and ultrastructural study of a series. Am J Surg Pathol. 1997;21:130-46
5. Chbani L, Guillou L, Terrier P, et al. Epithelioid sarcoma. Am J Clin Pathol. 2009;131:222-27
6. Ross HM, Lewis JJ, Woodruff JM, Brennan MF. Epithelioid sarcoma: Clinical behavior and prognostic factors of survival. Ann Surg Oncol. 1997;4:491-95
7. Panagiotopoulos N, Gelvez-Zapata S, Rassl D, et al. Primary epithelioid sarcoma of the pleura: An intricate diagnosis. Ann Thorac Surg. 2013;96:e79
8. Ouahbi H, Akasbi Y, Oualla K, et al. [Pleural epithelioid sarcoma, about a case with review of the literature.] Pan Afr Med J. 2016;25:120-27 [in French]
9. Setojima Y, Shimada Y, Tanaka T, et al. Prognostic impact of solid-part tumour volume doubling time in patients with radiological part-solid or solid lung cancer. Eur J Cardiorhorac Surg. 2019;57:763-70
10. Nakahashi K, Shiono S, Nakatsuka M, Endo M. Prediction of lymph node metastasis of clinical stage IA non-small cell lung cancer based on the tumor volume doubling time. Surg Today. 2022;52:1063-71
11. Schwartz M. A biomathematical approach to clinical tumor growth. Cancer. 1961;14:1272-94
12. Chan JKC, Galateau-Salle F, Nicholson AG, et al, editors. Tumours of the pleura and pericardium. In: WHO Classification of Tumours Editorial Board. WHO Classification of Tumours, 5th Ed., Vol.5 Thoracic Tumours. Lyon: IARC; 2021;193-226

Indexed in: [PMC] [PubMed] [Emerging Sources Citation Index (ESCI)] [Web of Science by Clarivate]